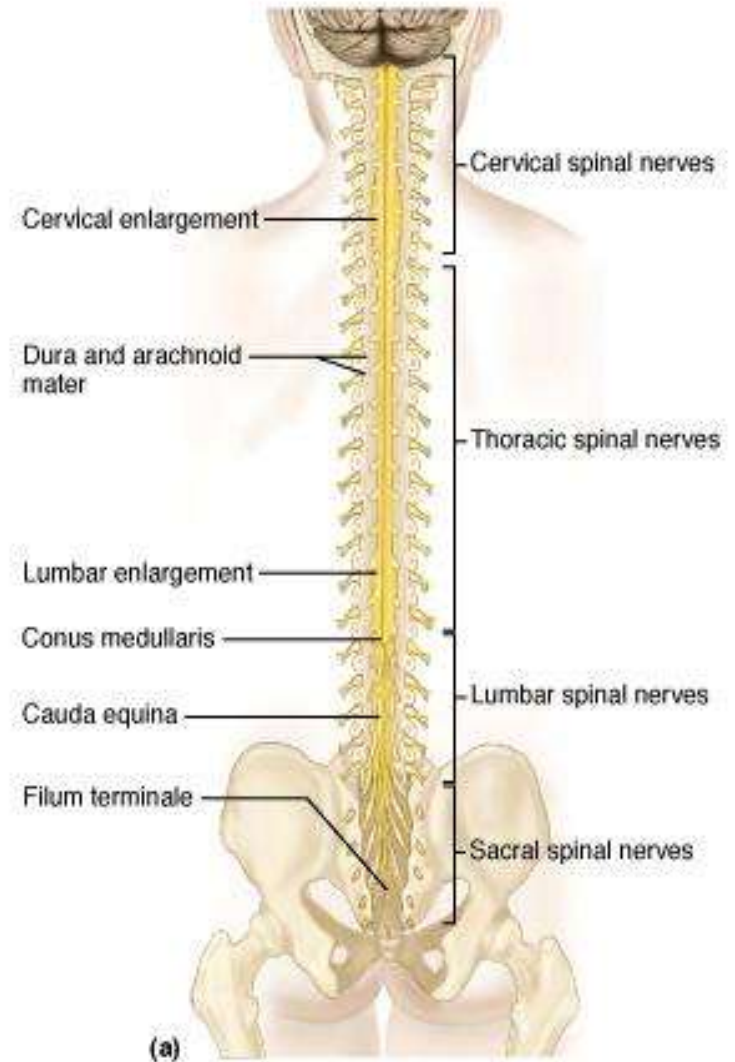
TUMORI SPINALI

Gruppo neurooncologico
21 novembre 2014

Spine and Spinal cord Tumors

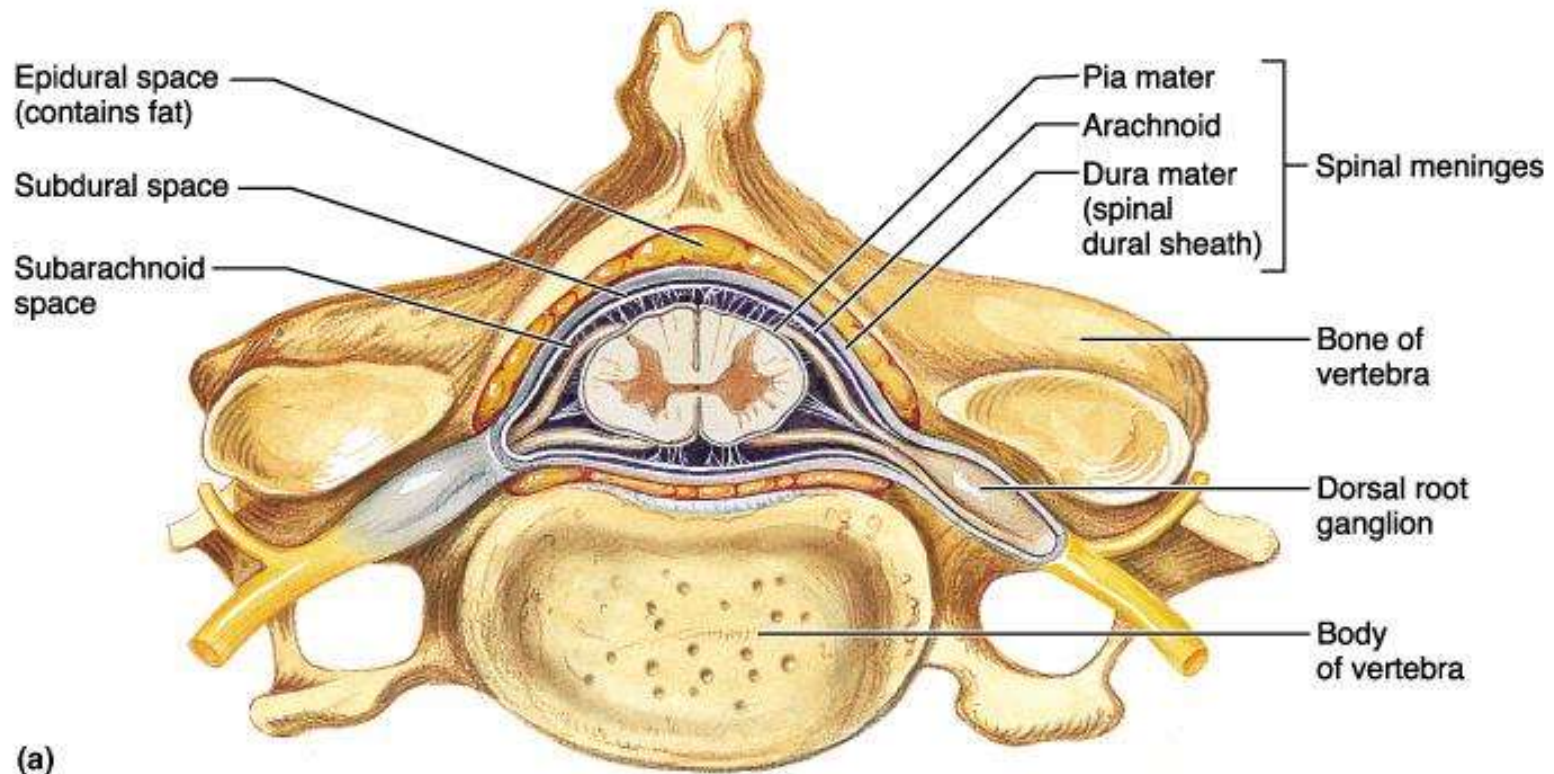
Tumori che originano nel midollo, nelle radici, nelle meningi o nelle vertebre, che causano compressione sul midollo e sulle radici nervose

Spine tumors

TUMORI DELLA COLONNA
benigni

- OSTEOCONDROMA 4%
- OSTEOMA OSTEOIDE 9%
- OSTEOBLASTOMA 10%
- EMANGIOMA 5%
- CISTI ANEURISMATICA 10%
- GRANULOMA EOSINOFILO 4%
- TUMORE A CELLULE GIGANTI 1-2%

Anatomy of the Spinal Cord

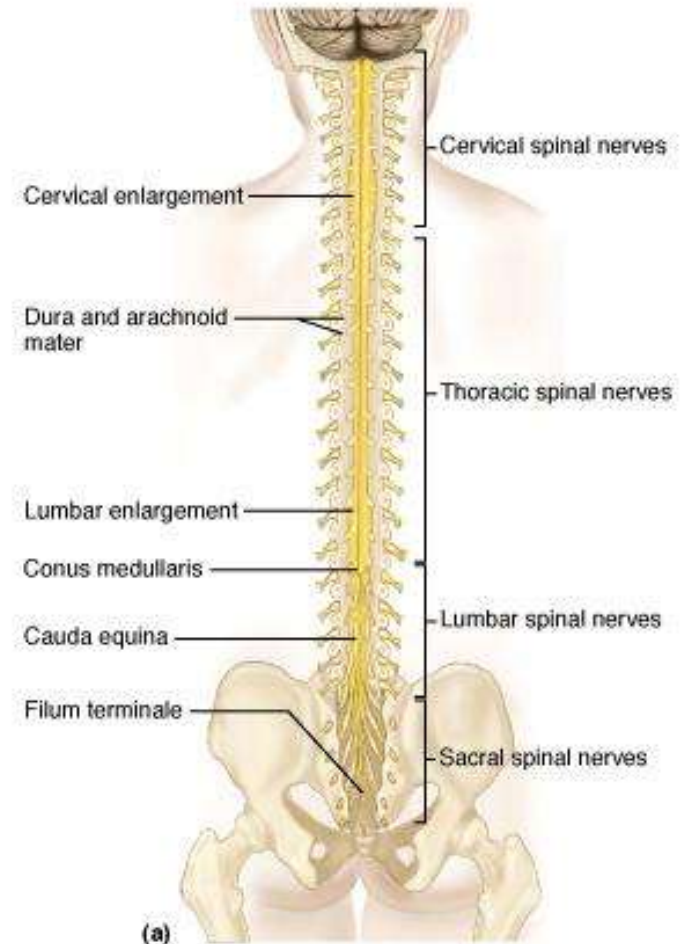
Figure 13.30a

Spine tumors

TUMORI DELLA COLONNA maligni

- MIELOMA/PLASMOCITOMA
- CORDOMA
- CONDROSARCOMA
- OSTEOSARCOMA (S. EWING)
- METASTASI (MAMMELLA-POLMONE-PROSTATA-RENE)

OGNI dolore rachideo della durata di oltre 6 settimane in bambini o in giovani adulti va indagata con TAC con mdc

TUMORI MIDOLLARI

I tumori midollari primitivi sono meno frequenti rispetto ai tumori cerebrali.
Circa 2/3 dei tumori primitivi midollari sono meningiomi e neurinomi.
Circa il 10% dei tumori spinali è intramidollare.

Anatomy of the Spinal Cord

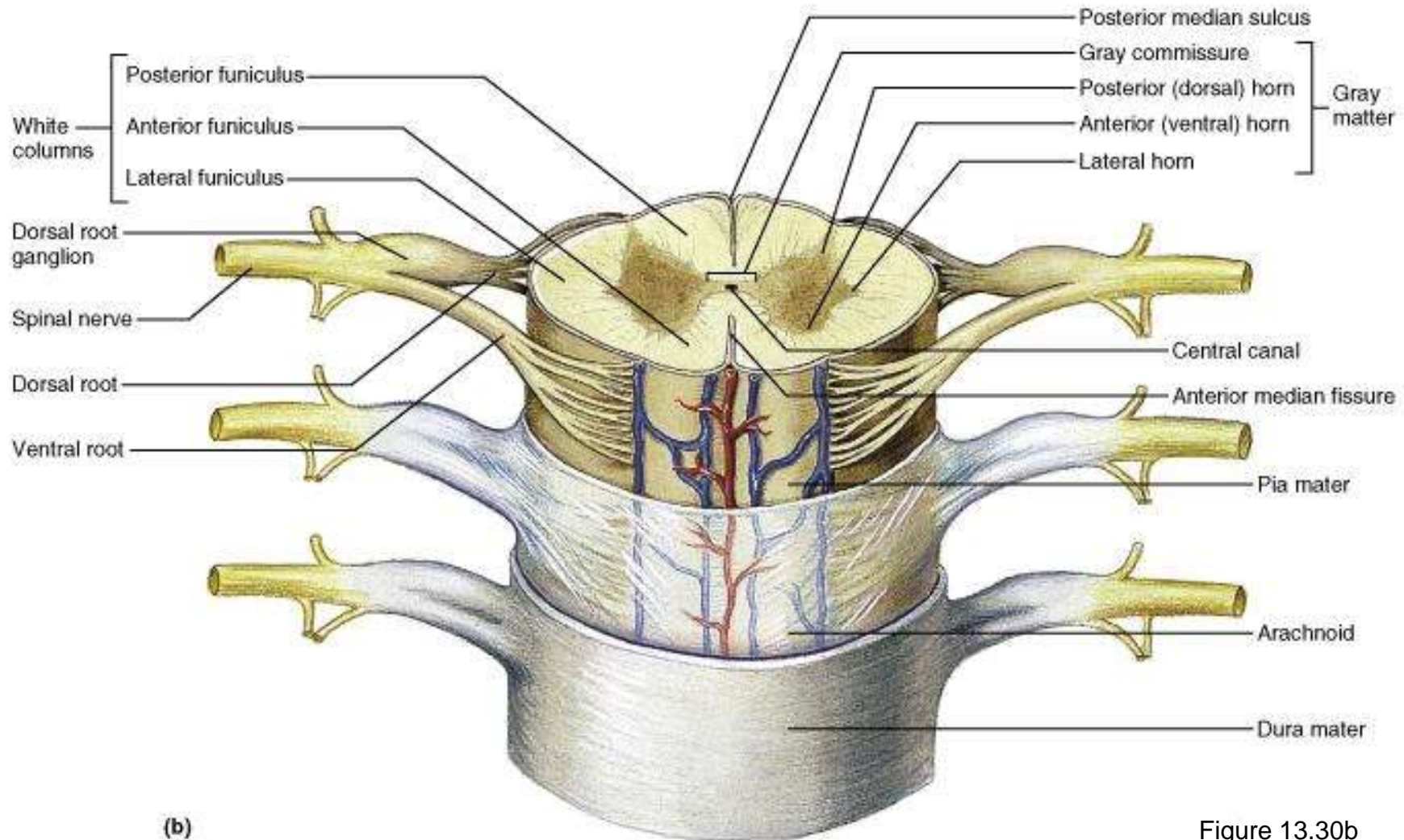



Figure 13.30b

INTRADURALI

INTRAMIDOLLARI :

EPENDIMOMI

ASTROCITOMI

CAVERNOMI

EMANGIOBLASTOMI

IUXTAMIDOLLARI:

SCHWANNOMI

NEUROFIBROMI

MENINGIOMI

SPINAL CORD TUMORS IN ADULTS

Extramedullary (2/3 of cases)	
Nerve sheath tumor	(40%)
Meningioma	(40%)
Filum terminal ependymoma	(15%)
Miscellaneous * (rare)	(5%)
Intramedullary (1/3 of cases)	
Ependymoma	(45%)
Astrocytoma †	(40%)
Hemangioblastoma	(5%)
Miscellansous †	(10%)

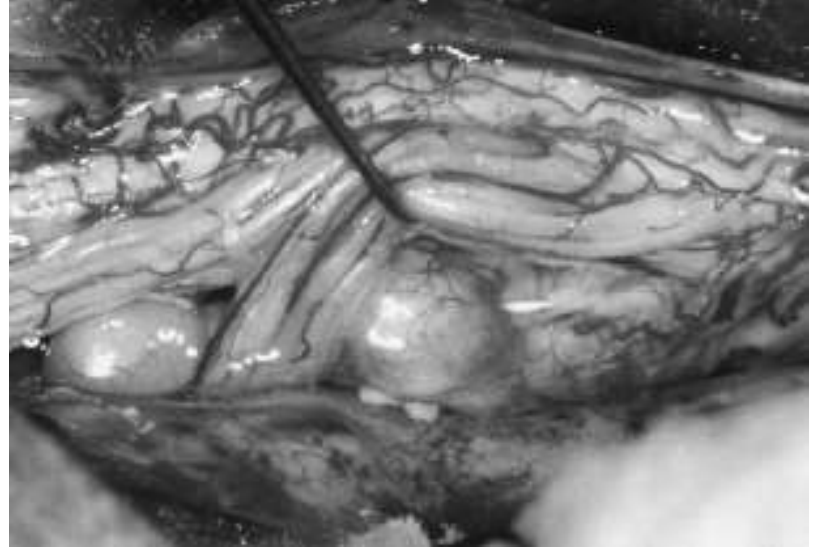
*Includes paraganglioma, drop metatasis, and ganglioma.

† includes oligodendroglioma, ganglioma, neurocytoma, and subependymoma.

†includes metastatic tumor. Inclusion tumor (e.g., lipoma), inflammatory pathology (e.g., abscess, tuberculoma, sarcoid), and vascular pathology (e.g., cavernous malformation, aneurysm).

sintomi

I **tumori intradurali extramidollari** (p. es., gli schwannomi, i meningiomi) causano dolore segmentale, con evoluzione verso il deficit segmentale di forza e infine paraparesi.
Neoplasie intramidollari: si tratta di gliomi ed ependimomi che frequentemente si estendono lungo parecchi segmenti spinali, simulando la siringomielia. Insorgono paraparesi progressiva, perdita delle sensibilità e deficit sfinteriale. Un tumore localizzato a un segmento midollare può simulare una neoplasia extramidollare, ma il dolore è meno evidente e i deficit sfinteriali sono più precoci.

- **Tumori della guaina dei nervi 40% of E.M.S.C.T**
  - **Schwannomi**
    - » **Dalle cellule di Schwann**
  - **Neurofibromi**
    - » **Da cellule di Schwann, cellule del perinevrio e da fibroblasti.**
- **Neurofibromi e schwannomi hanno caratteristiche demografiche, istologiche e biologiche differenti.**

- Neurofibromi
  - Aspetto istologico
    - Abbobdanza di tessuto fibroso
    - Evidenza di fibre nervose nello stroma tumorale.
    - Ingrossamento fusiforme (plessiforme) del nervo interessato.

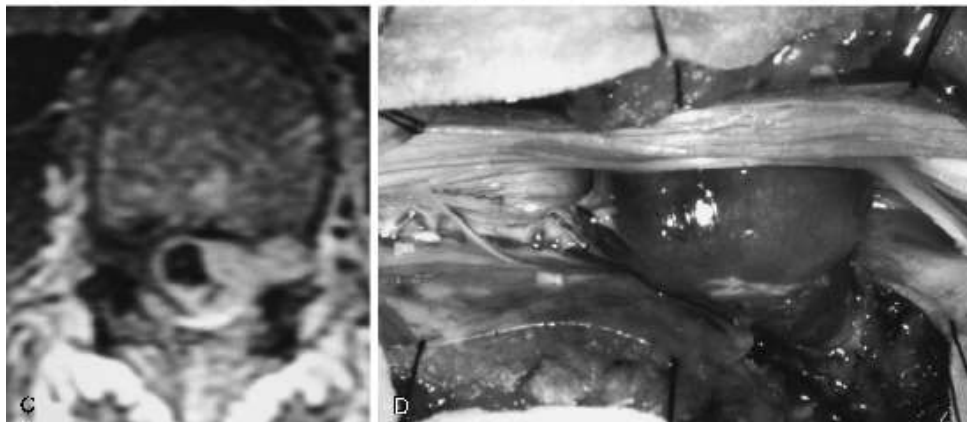
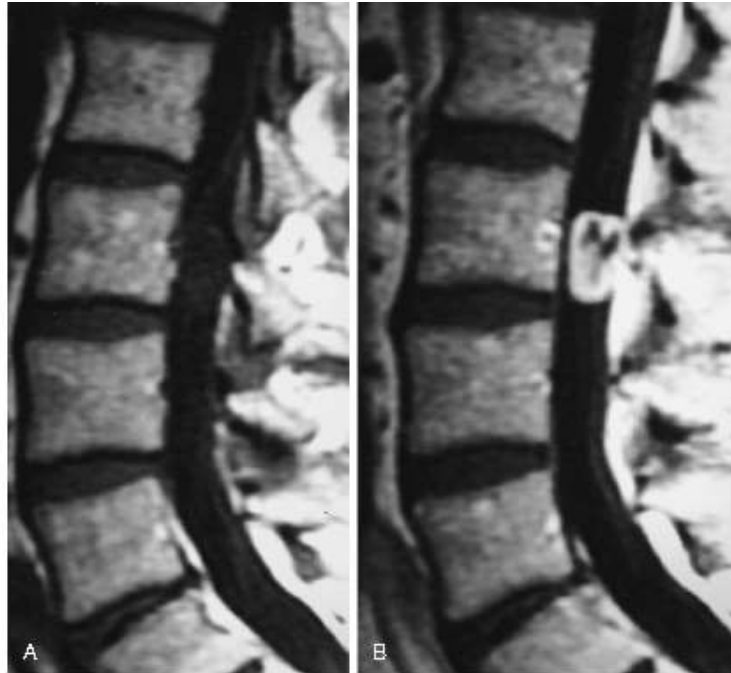
- Neurofibromi multipli fanno porre diagnosi di Neurofibromatosi:

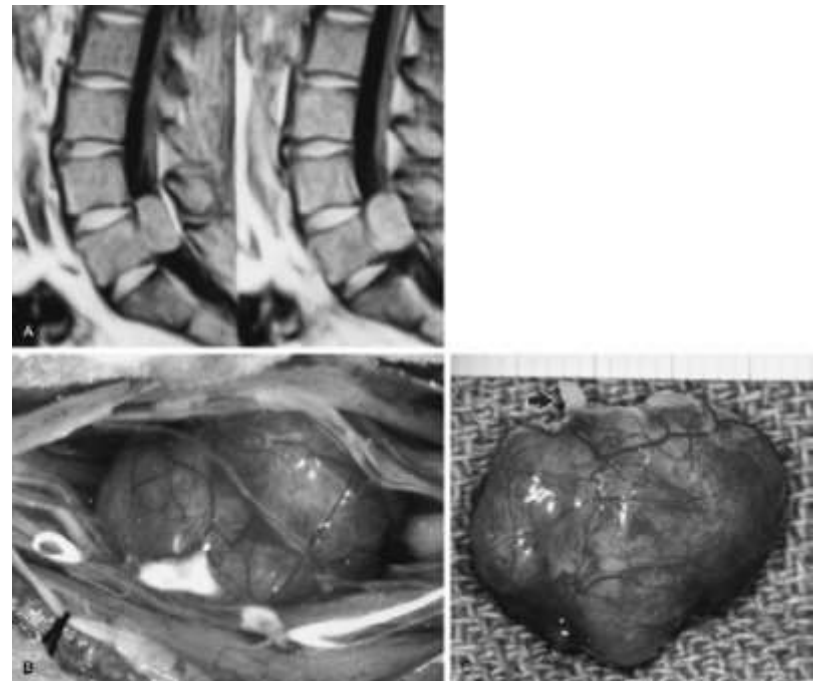
Tipo I più frequente – associata a fibromi cutanei, macchie caffè e latte, anomalie scheletriche e localizzazioni a livello di nervi spinali (spesso asintomatica)

Tipo II più spesso associata a neurinoma dell VIII nc

Tumori della guaina neurale

- Maggioranza sono schwannomi solitari
- Picco di incidenza; 4^{th} – 6^{th} decade
- Maschi = femmine
- Maggioranza nasce dalle radici dorsali
- I Neurofibromi nascono più spesso dalle radici ventrali.
- 10% epidurali o paraspinali.
- 1% intramidollari
- Raramente maligni quando lo sono sopravvivenza difficilmente superiore ad 1 anno.

A
B
C
D

Terapia chirurgica

- 1) rimozione totale o subtotale
  - Recidiva rara, più spesso in quelli a clessidra

Meningiomas

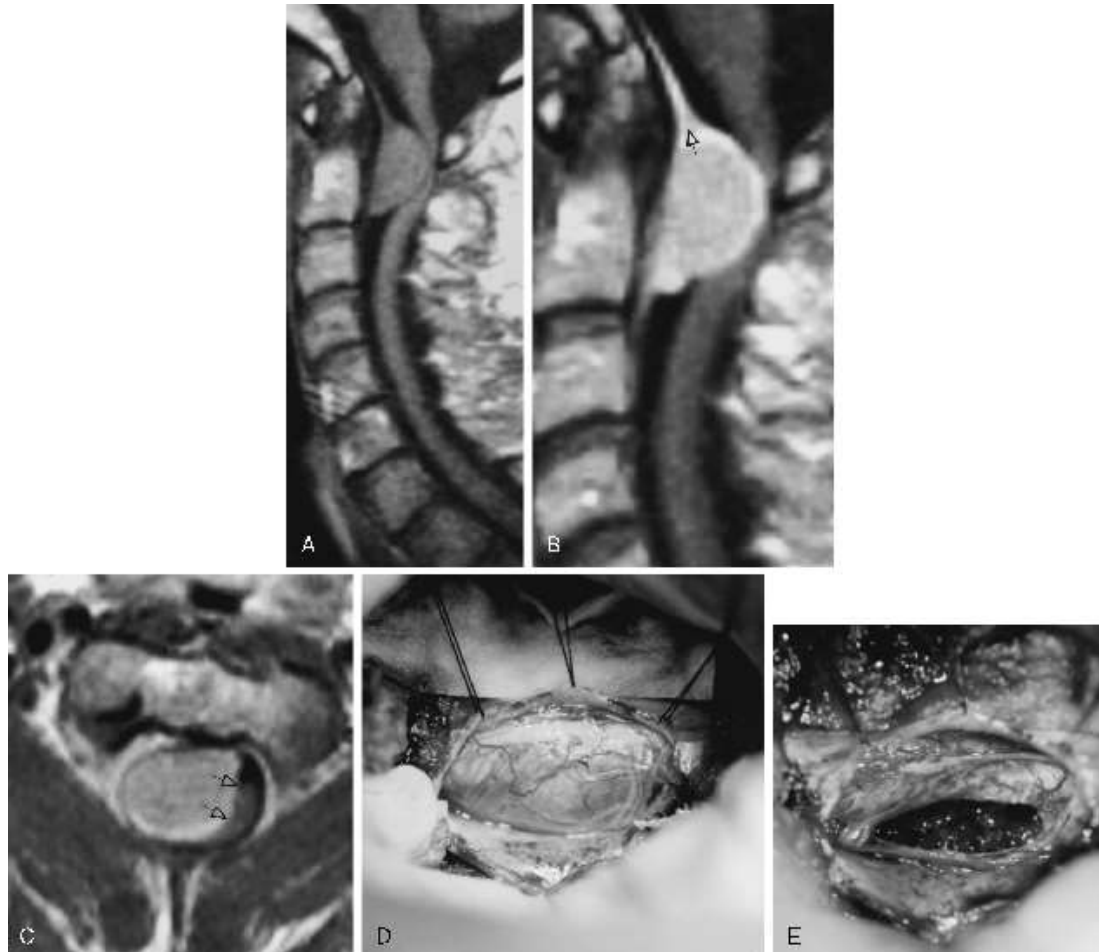
- Meningiomas= nerve sheath tumors= 40% of E.M.S.C.T
- Arise from: (mesodermal origin)
- Arachnoid cap cells
- embedded in the dura near the root sleeve
- Pia
- Dural fibroblast
- Peak incidence= 60-80 year, 75 -80% occur in women 80% are **thoracic**
- Upper cervical and foramen magnum are other common sites ventral position
- Ventrolateral position

Meningiomas

- 90% intradural
- 10% intra-extradural & entirely extradural
- gross characteristic smooth fibrous
- Histological characteristic :

    Broad dura attachment

    Bony involvement does not occurred

Meningiomas

(Meningiomas)
Treatment 1

- complete surgical removal= treatment of choice
- Favorable features compared to intracranial meningiomas
- Absence of bony involvement
- Lack of venous sinus or major blood vessel involvement

10-15% recurrence rate of intraspinal meningiomas at 10 years after total or near total removal.

Meningiomas

Treatment 2

- **technique:**
- 1. Standard posterior laminectomy
- 2. Unilateral laminectomy and facetectomy
- 3. Costotransversectomy for ventral thoracic tumors
- 4. Lateral extra cavitary for ventral thoracic tumor
- 5. Extreme lateral approach (Sen & Sekhar method) for significant tumor components above foramen magnum.

Meningiomas

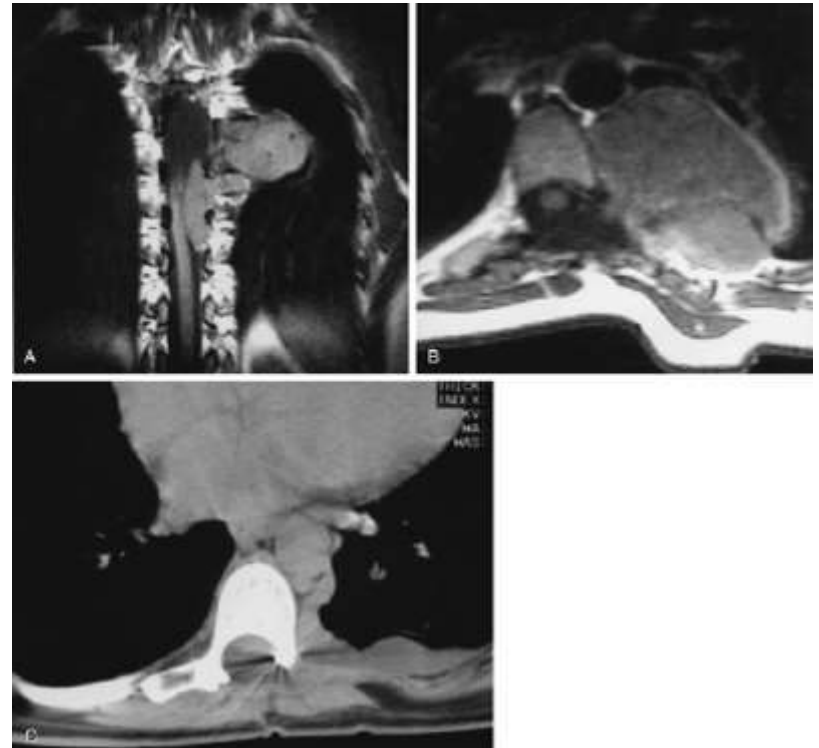
Treatment 3

- 6. large tumors are debulked and then removed, dural attachement is excised or extensively coagulated.
- 7. devide archnoid adhesions to prevent
  - A. Tethering
  - B. delayed syrinx formation
  - C. arachnoiditis
  - D. hydrocephalus

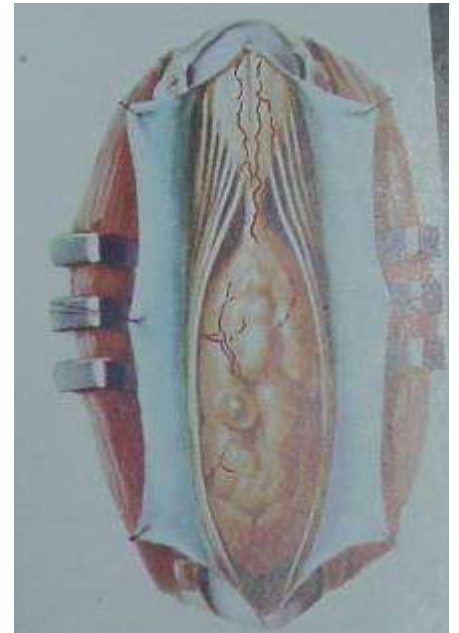
Meningiomas

Treatment 4

- Rarery spinal meningiomas may extend through a dural nerve root sleeve & present as dumbbell tumor

- Management
  - Excision and patch graft
  - Extensive in situ coagulation

FILUM TERMINAL EPENDYMOMA

- 40% of spinal canal ependymomas arise within filum terminale
- **Proximal portion of filum = most common site**.

  Astrocytoma, oligodendroglioma and para ganglioma may also originate in the filum terminal
- 3^{th} – 5^{th} decade= most common
- Men= women
- Myxopapillary= most common
- Almost all are biologically benign

Filum Terminale Treatment

- Size and relationship of tumor to the surrounding roots and cauda equina determine the role of surgery.
- Gross total en bloc resection should be attempted whenever possible.
  - For small and moderate size tumor
  - Well circumscribed.
  - Inter and decompression may increase the risk of dissemination .
  - Recurrence after en bloc resection are rare.

Filum ependymomas treatment

- Large filum terminal ependymomas can present significant problems for surgical resection.
- High risk of seminarian through CSF (have been present for many years)
- Entire neuraxis should be evaluated.
- These tumor may reach enormous size.
- May insinuate among the roots and within the arachnoid sheaths of the cauda equina

Filum ependymomas treatment

- **SurgicalTechnique**

- piecemeal & subtotal removal
- Diminish the tumor build
- 20% recurrence rate if a near total piecemeal removal has been acheaved.
- **Radiotherapy**
- delay in situation in which surgery can be contem plate
- For early tumor recurrence in biologically aggressive tumors.
- Postoperatively: for significant tumor remnance or dissemination
- After repeated surgery for tumor recurrence.

Tumori intramidollari

- **primitivi gliali**

  80%

Tumori intramidollari

DD.lesioni non neoplastiche

- Infiammatorie
- Ascessi batterici
- Mieliti
- Sclerosi multipla
- Tuberculoma
- Sarcoidosi

ASTROCITOMI MIDOLLARI

- Sono il 3% di tutti gli astrocitomi
- 60 % regione cervicale – passaggio cervicotoracico
- Maggior frequenza nelle prime tre decadi (il 90% nella prima decade)

ASTROCITOMI MIDOLLARI

Sono un gruppo di tumori eterogeno dal punto di vista della:

-istologia
-caratteristiche macroscopiche
-biologia
-storia naturale

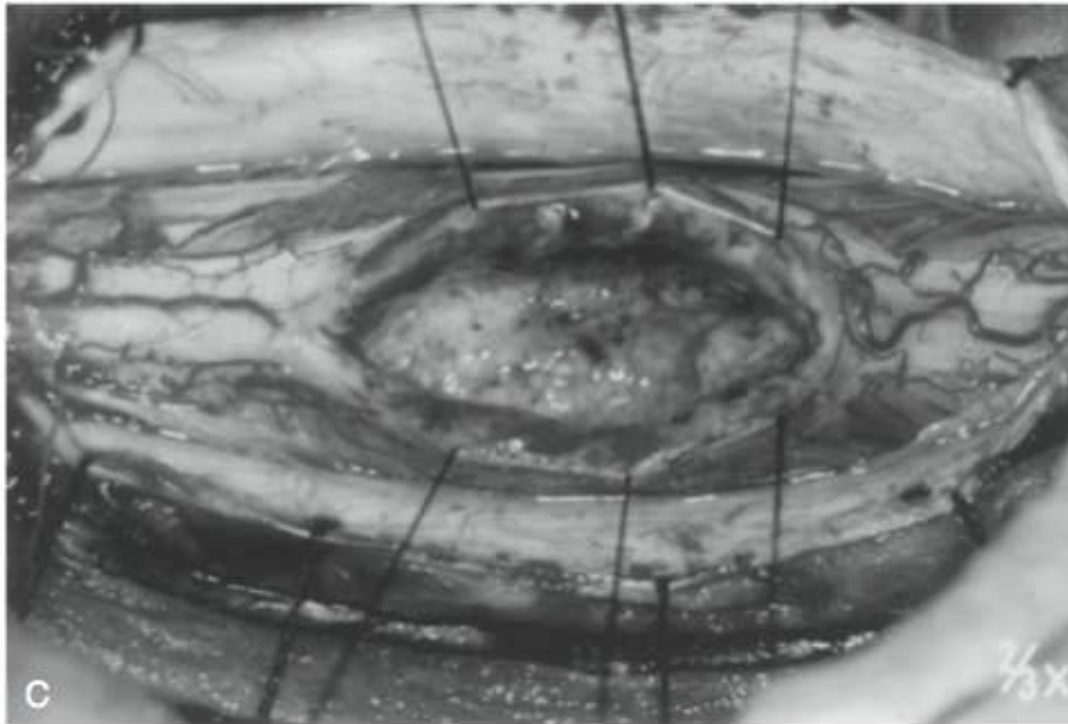
Astrocitomi intramidollari

- Include
  - Astrocitomi fibrillari di basso grado
  - Astrocitomi pilocitici
  - Astrocitomi maligni
  - Gangliogliomi
  - Oligodendrogliomi

    - 90% degli astrocitomi pediatrici sono benigni
    - 10% dei gliomi pediatrici sono astrocitomi maligno o glioblastomimaligni are malignant astrocytomas or glioblastomas

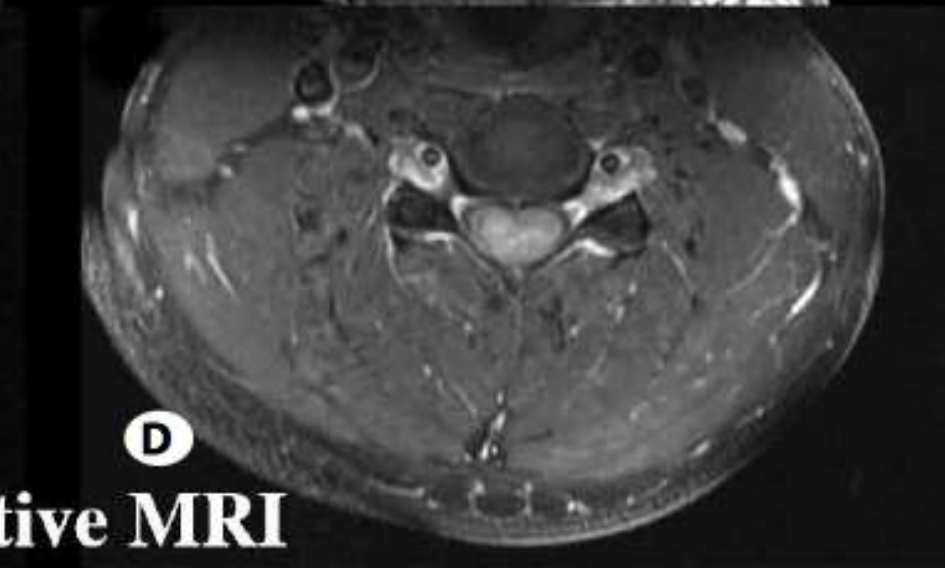
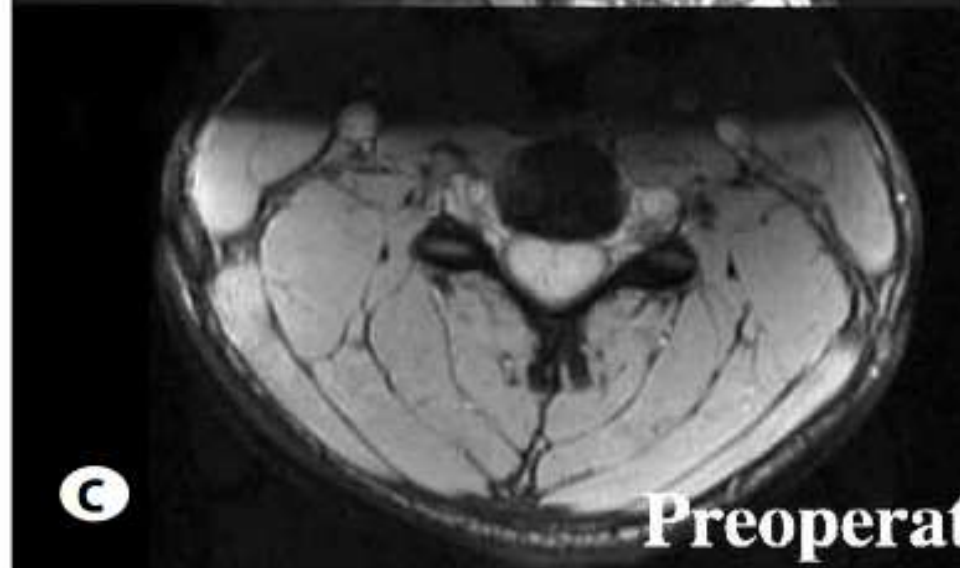
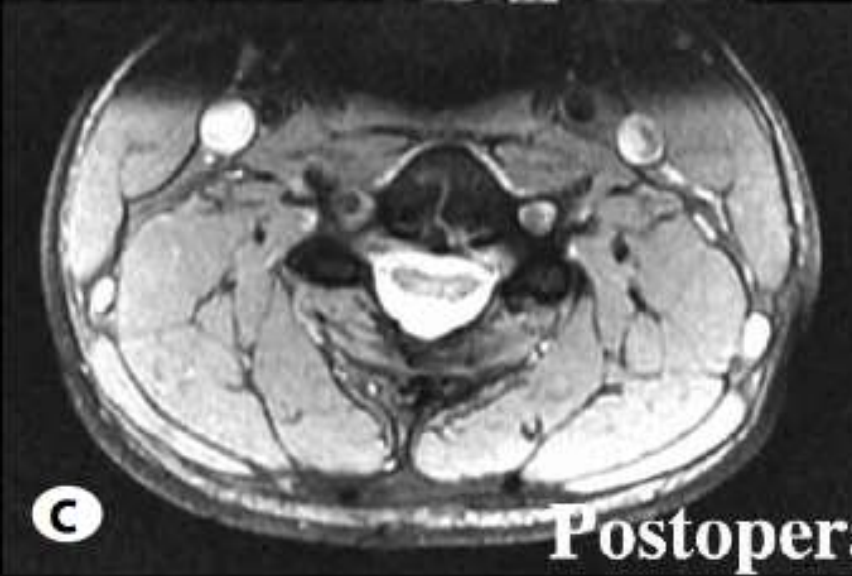
Tumori intramidollari

astrocitomi

- Gli astrocitomi fibrillari prevalgono nell’adulto
- L’astrocitoma pilocitivo prevale nell’adolescenza
- Nell’adulto il 25% degli astrocitomi sono maligni.

IGURE 101-2. Gadolinium-enhanced sagittal (**A**) and (**B**) axial MRI scans show a focal, uniformly enhancing intramedullary mass. Irregu
ımor margins are apparent on the axial view. At surgery, a focal benign astrocytoma with infiltrating margins was radically, but sub
ılly, resected. An intraoperative photograph after tumor resection (**C**) demonstrates indistinct infiltrating tissue margins.

Tumori intramidollari

Ependimoma

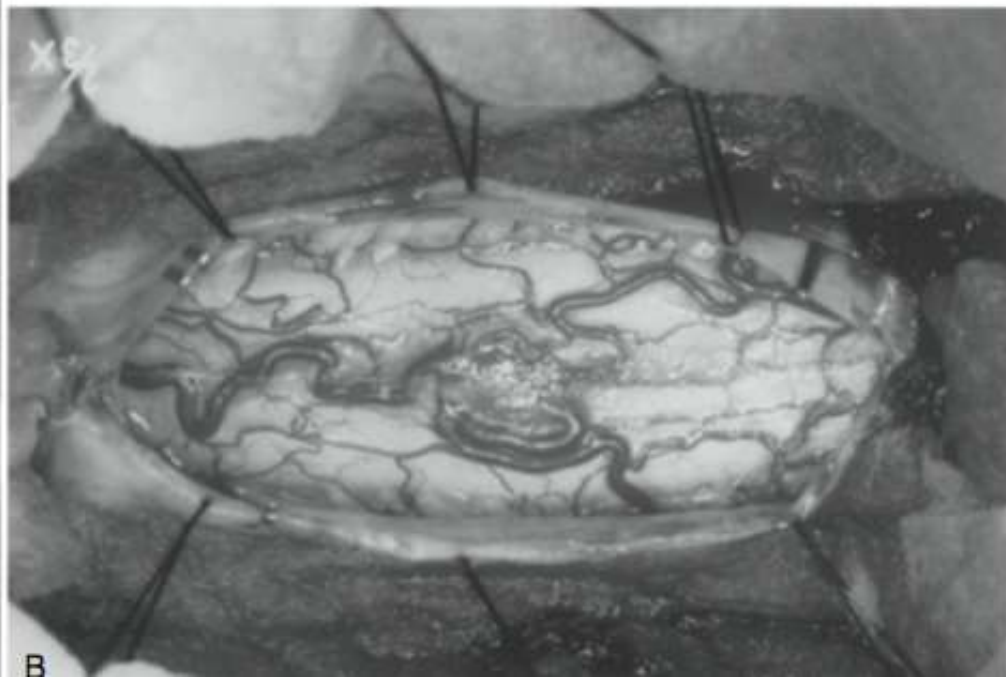
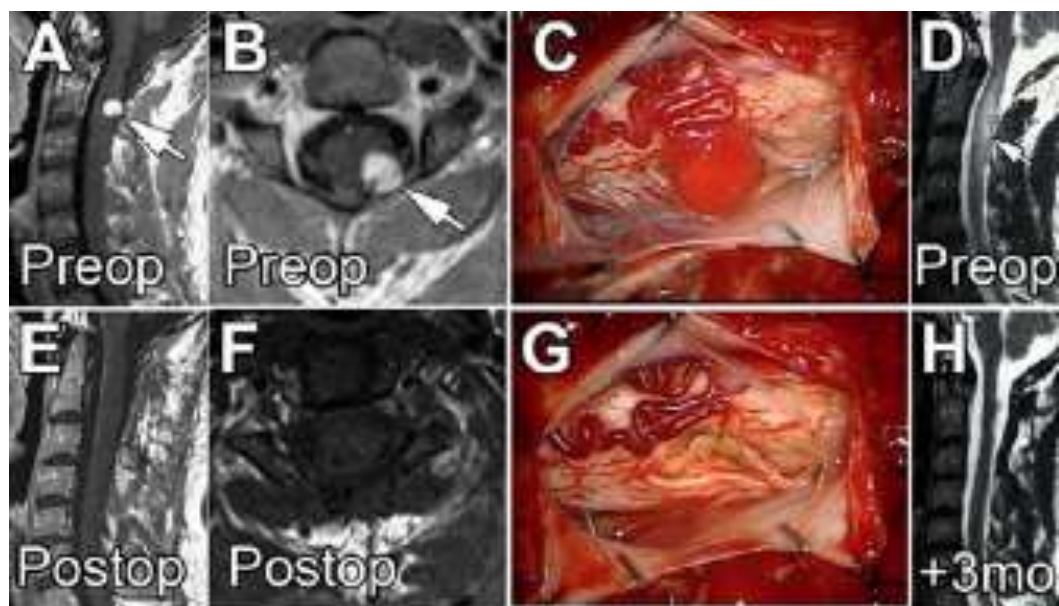
- Il più frequente tumore IM nell’adulto.
- Nessuna prevalenza di sesso.
- Molti sottotipi istologici.(praticamente tutti benigni)
- Privi di capsula ma ben circoscritti. Origina da cellule gliali.

12.8 mm
MRI TOTAL SPINE
C1
C2
C3
C4
C5
C6
C7
50 mm
A
71.1 mm
B
C
D
Preoperative MRI

Postoperative MRI

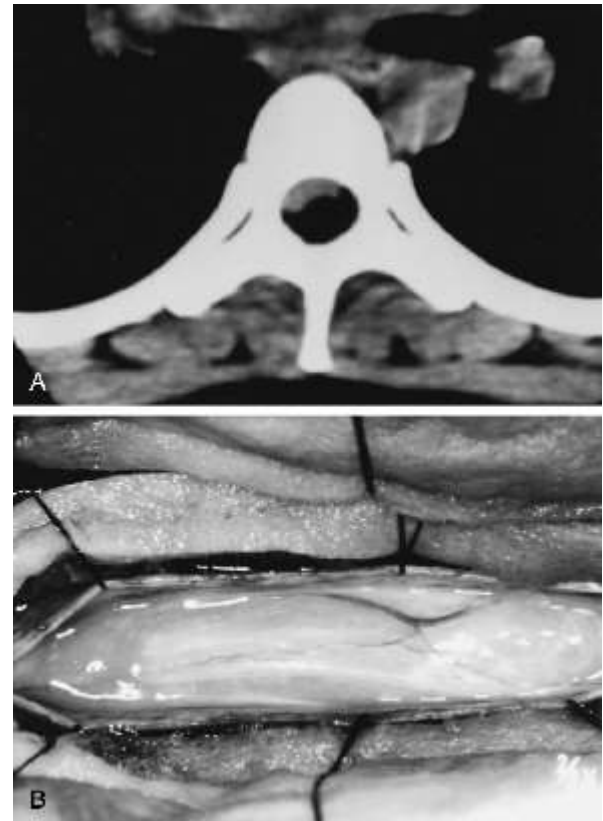
Tumori Intramidollari

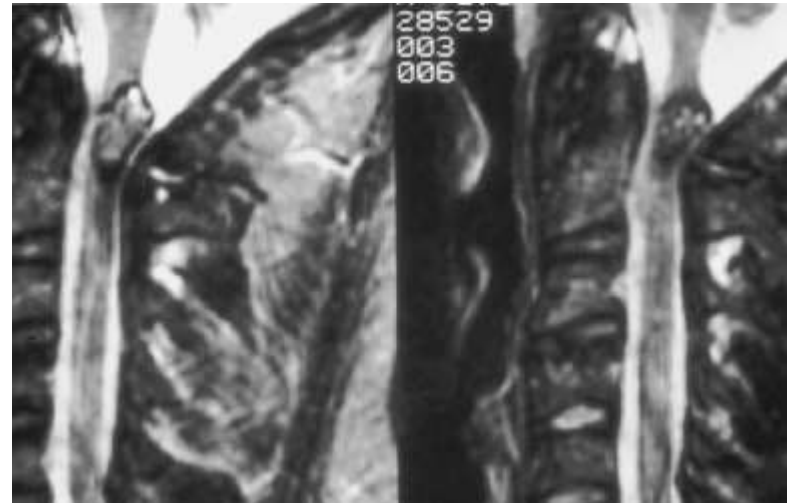
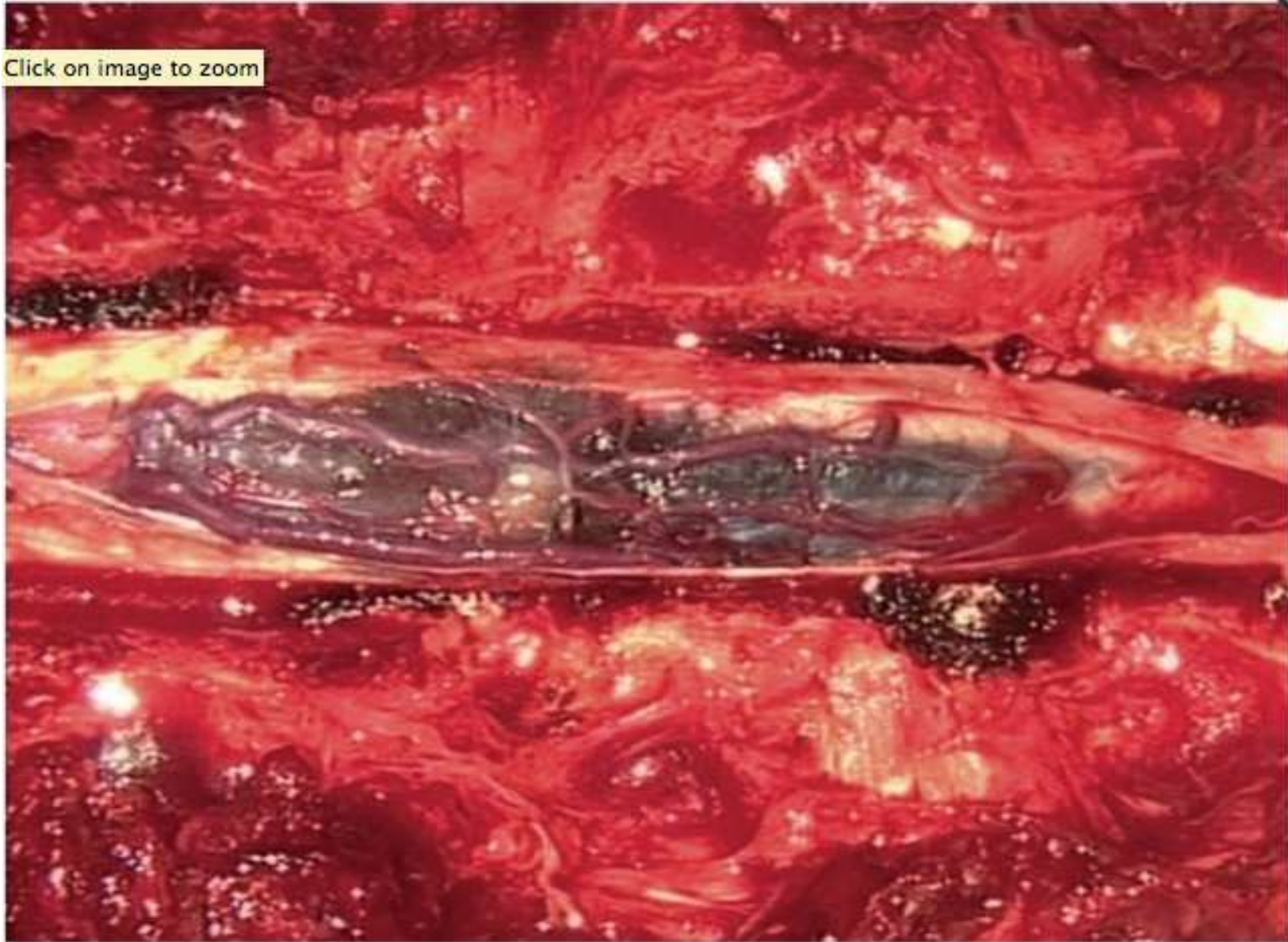
Emangioblastoma

- 3-8% di tutti i T.I.
- 15-25% in associatione con S. di von Hippel-Lindau.
- Rare in childhood.
- Tumori benigni di origine vascolare.
- Nettamente circoscritti non capsulati.
- Aderenza con la pia
- Localizzati prevalentemente in sede dorsale o dorsolaterale

A
B

A
Preop
B
Preop
C
D
Preop
E
Postop
F
Postop
G
H
+3mo

Tumori intramidollari

- Lesioni disembriogenetiche
  - Lipomi= I più comuni 1% di tutti I TI

Tumori intramidollari

- metastasi 2% (difficile diffusione ematogena)
  - Polmone mammella
  - Melanocitoma
- Malformazioni vascolari (cavernoma)

Operation microscope view of the surgical area. Dark gray pigmented intramedullary lesion is exposed after dural incision.

TUMORI INTRAMIDOLLARI CARATTERISTICHE CLINICHE

- Variabili
- Sintomi precoci aspecifici
- Durata dei sintomi prima della diagnosi lunga:

- 2-3 anni.
- Settimane o mesi in caso di lesioni maligne o metastatiche
- Decorso più breve se emorragie intratumorali (es ependimomi)

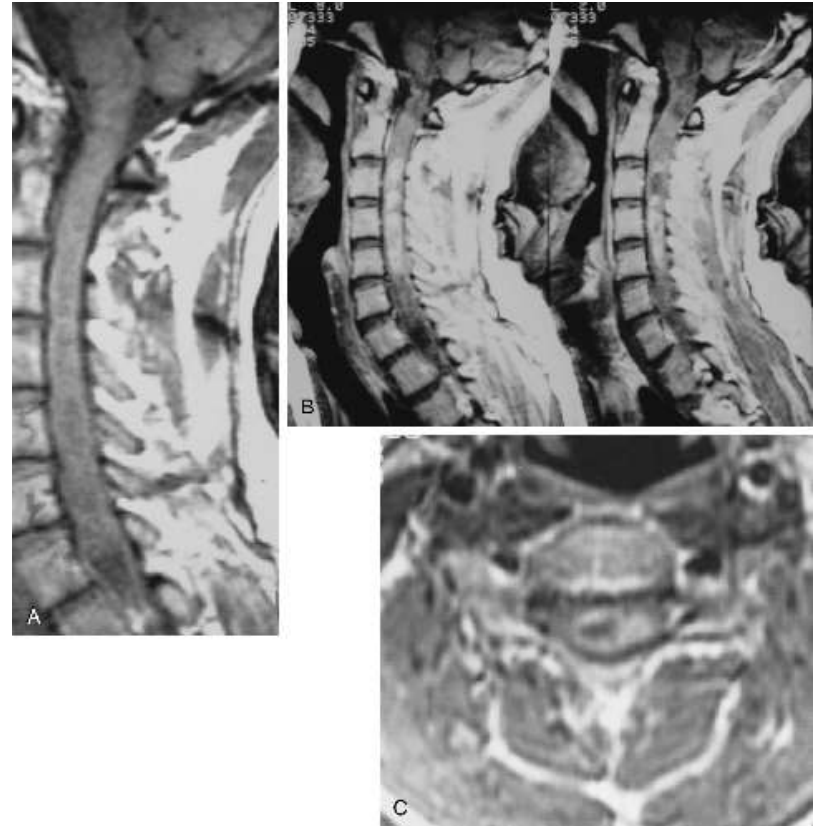
TUMORI INTRAMIDOLLARI

Caratteristiche cliniche

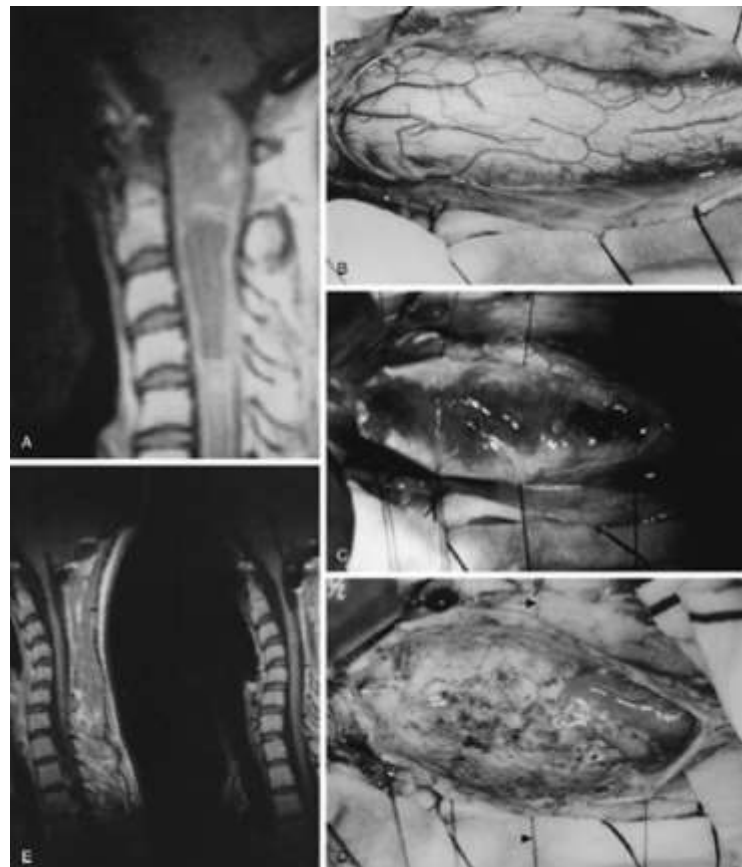
- Sintomi
  - Dolore = il più frequente
  - Disturbi sensitivi = 1/3 dei pazienti
  - Disturbi motori = 1/3 dei pazienti
  - Disestesie
  - Senso di addormentamento
  - -
  - -

Tumori Intramidollari

Radiologia e imaging

- Radiografia standard: obsoleta
- C.T Scan - Mielografia
- RM: esame di scelta
  - Molti tumori intramidollari sono isointensi o leggermente ipointensi in T1
  - Spesso si evidenzia solo un mal definito rigonfiamento del midollo nelle immagini in T1

Tumori intramidollari

aspetto radiologico

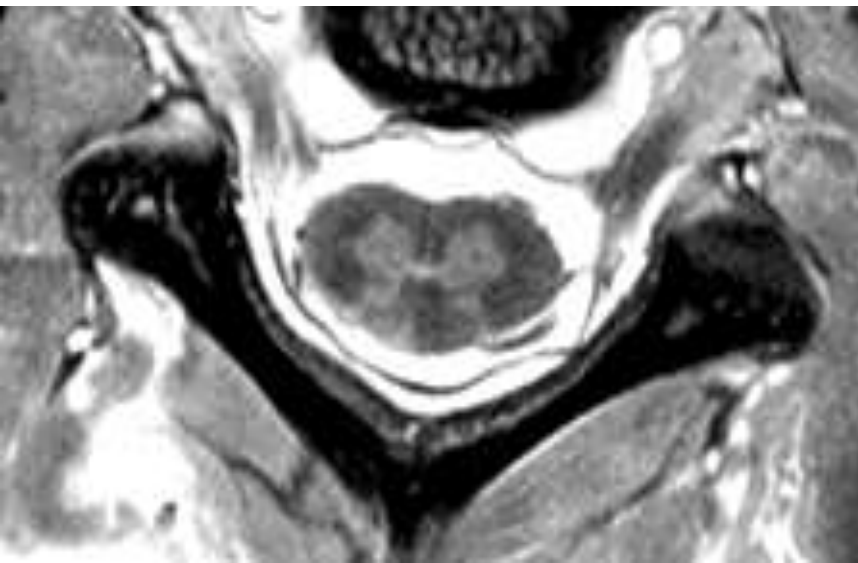
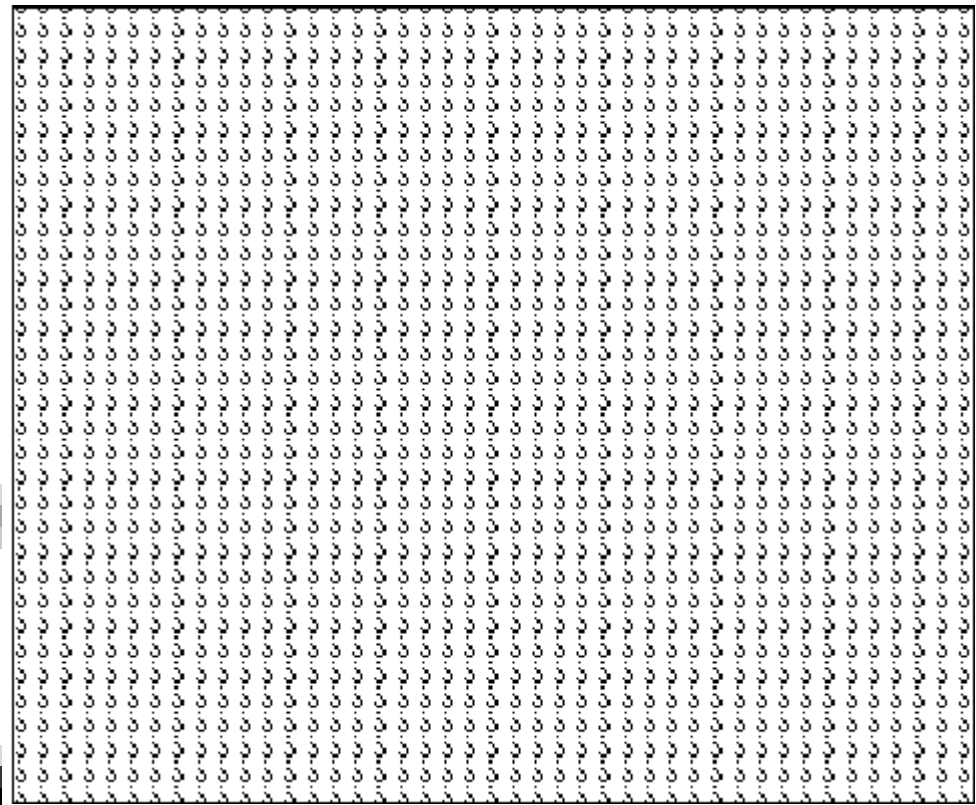
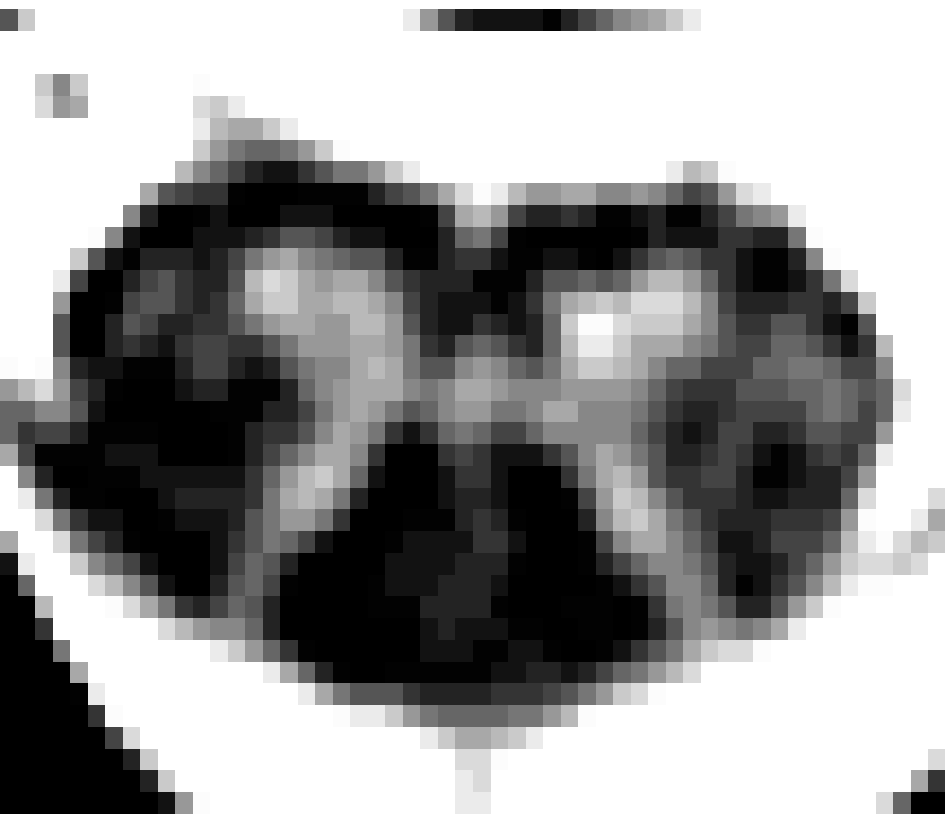
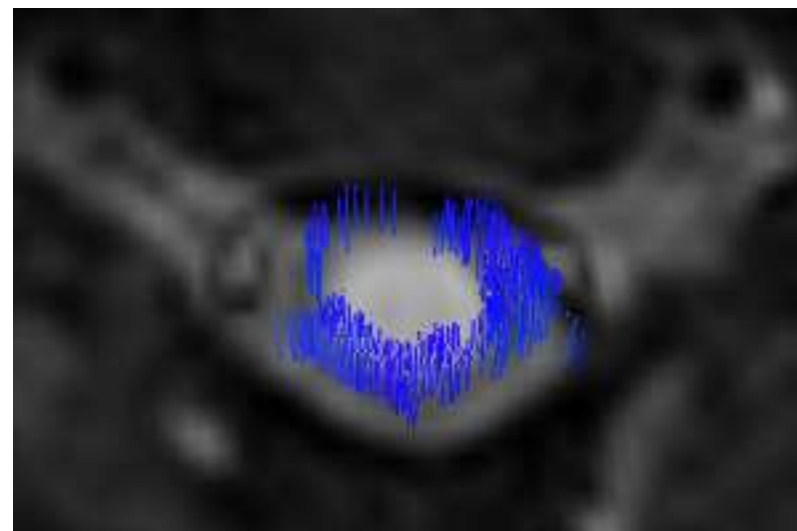
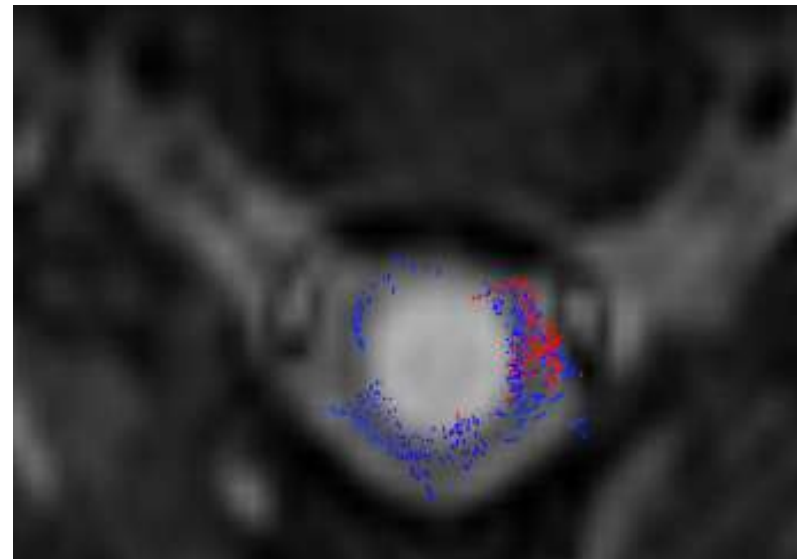
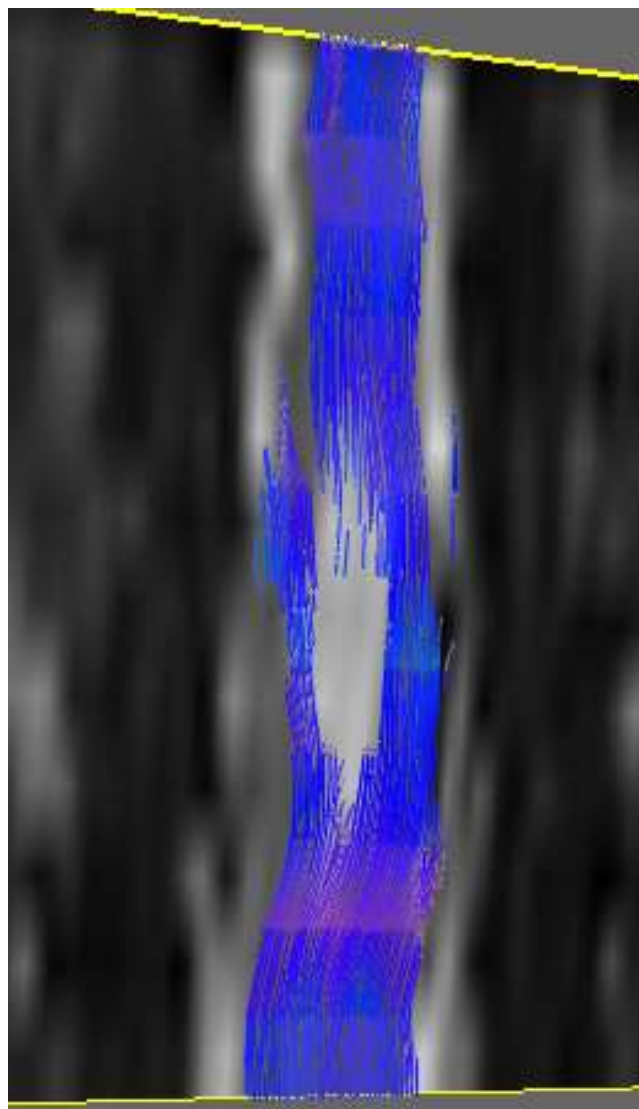
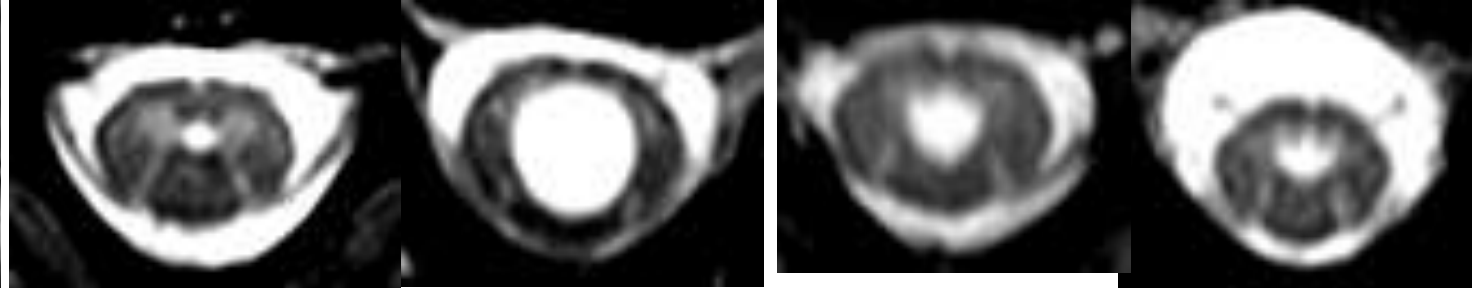
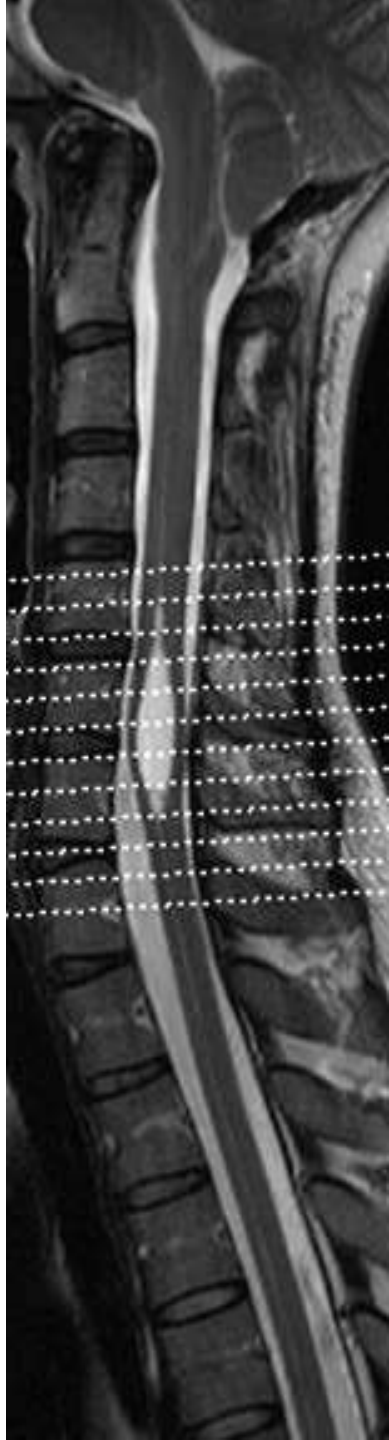
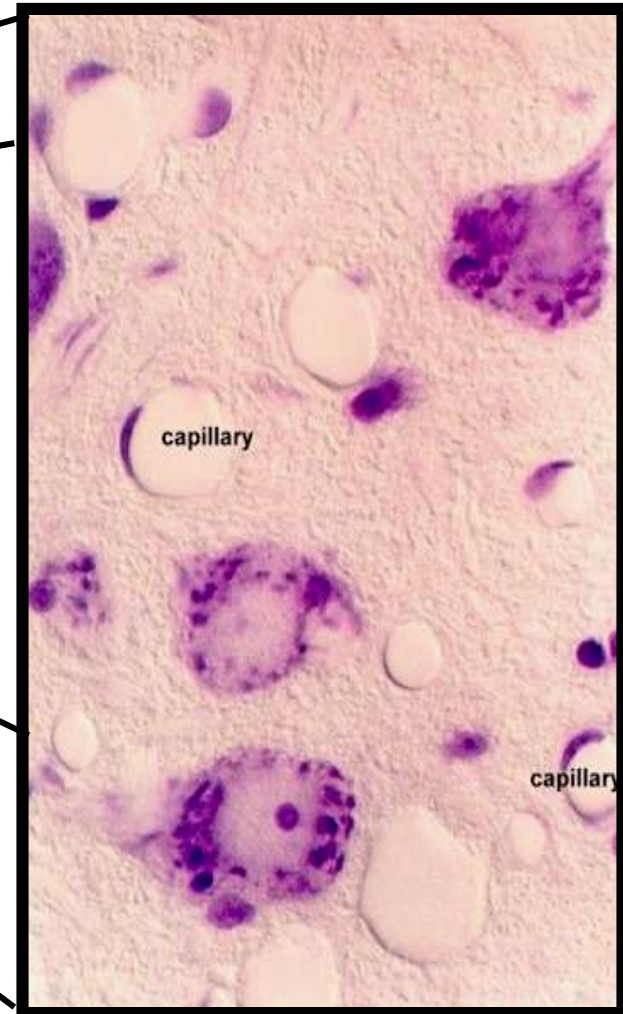
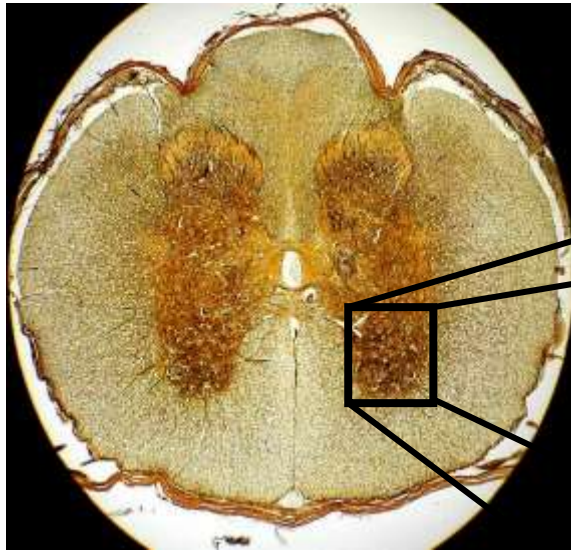
- Più sensibili le immagini pesate in T2 (molti tumori sono ipointensi in T2)
- Le immagini più specifiche si hanno in T2 con mdc in quanto quasi tutti I TI assumono contrasto
  - Assunzione uniforme del contrasto (ependimomi)
  - Spesso cisti polari
  - Aspetto variegato (astrocitomi)
- la diagnosi istologica non può essere fatta sul neuroimaging.

Imaging of Spinal Cord Function and Physiology

Spyros S. Kollias
University of Zurich, Switzerland

USZ / NRA

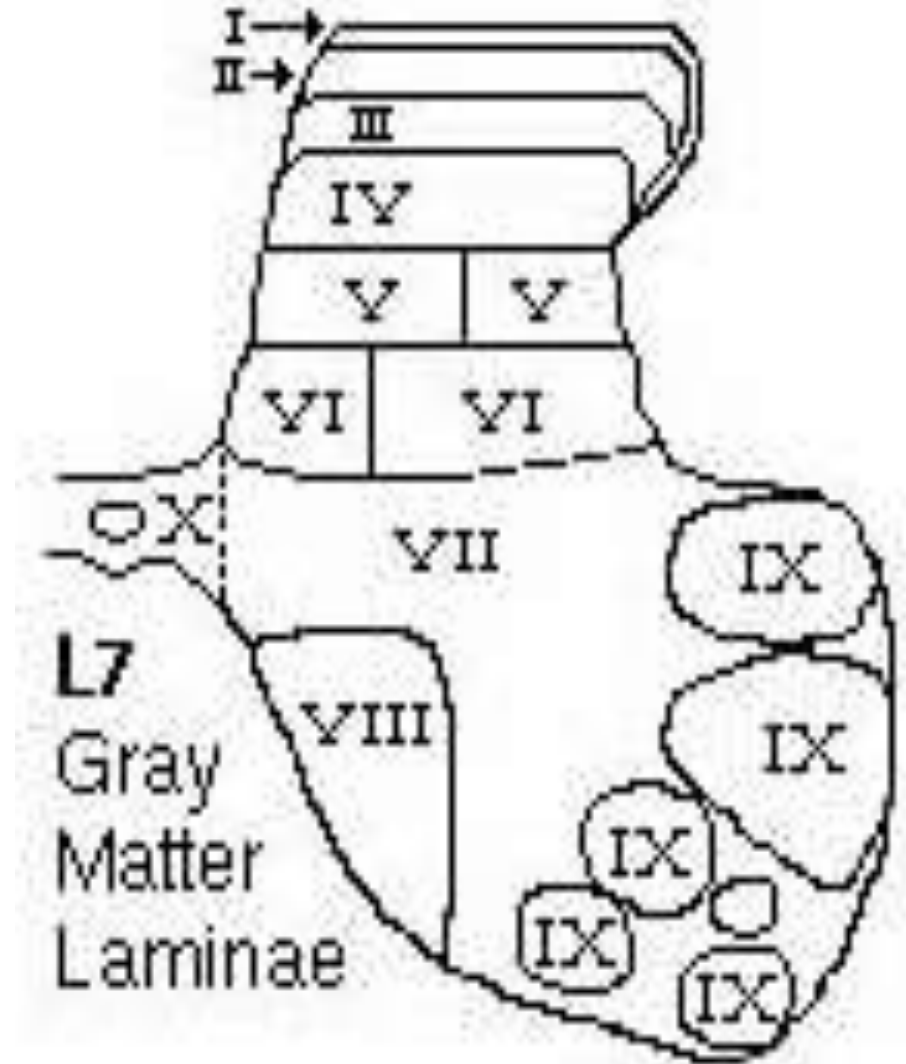
Research focus

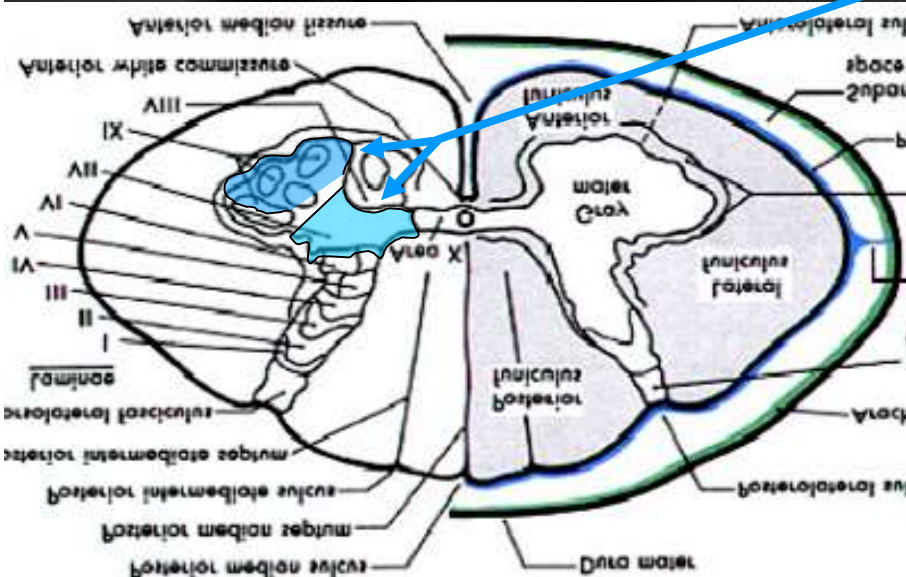
- Optimization of the techniques:
  - High-resolution morphological MRI
  - Diffusion-tensor MRI
  - Functional MRI
  - MR Spectroscopy

High-Resolution Morphological Imaging

White matter fiber tracts (bundles)

Functional MRI of the Spinal Cord

- <u>gray matter:</u>
- high density neuron cell bodies
- high capillary density

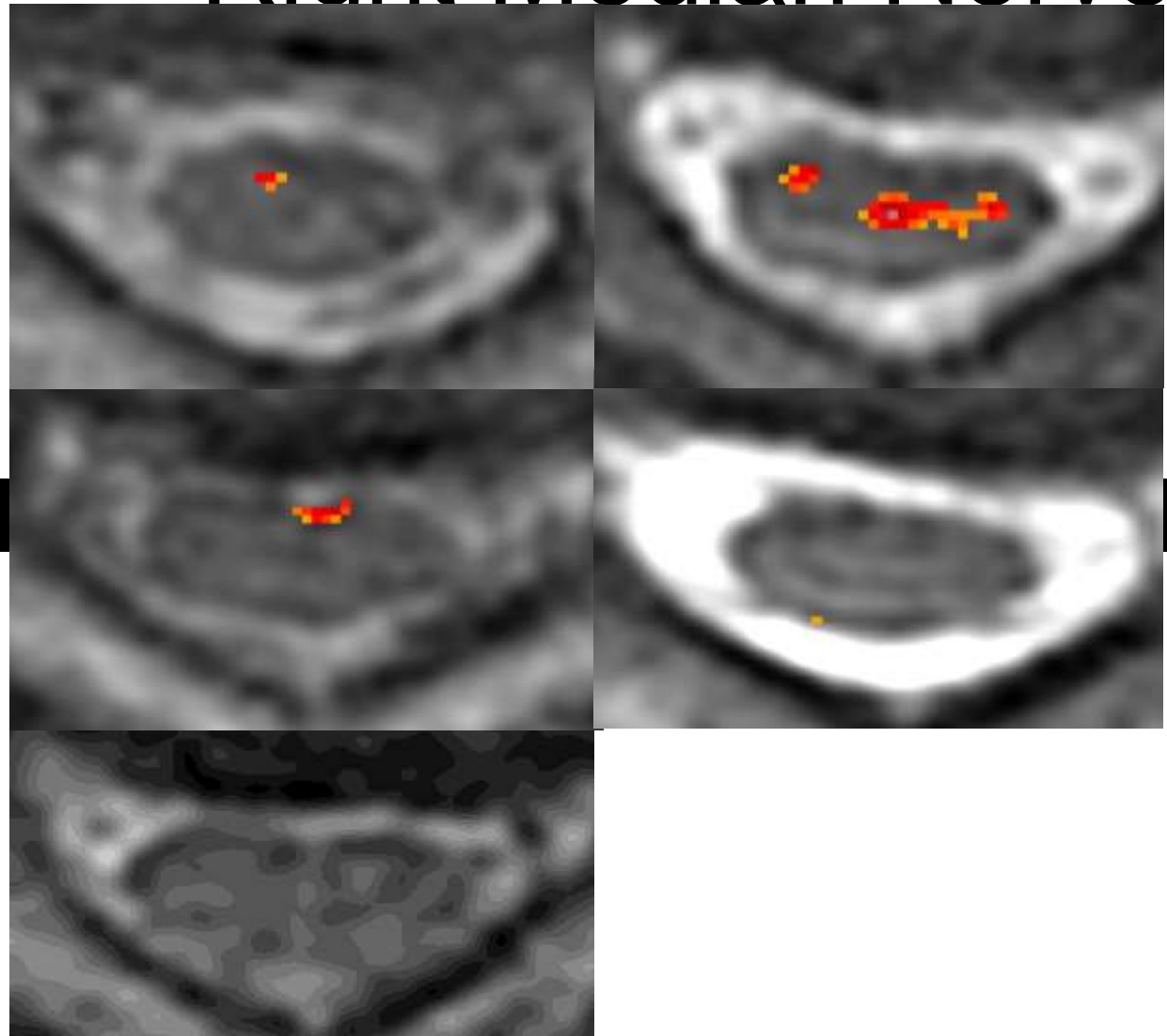
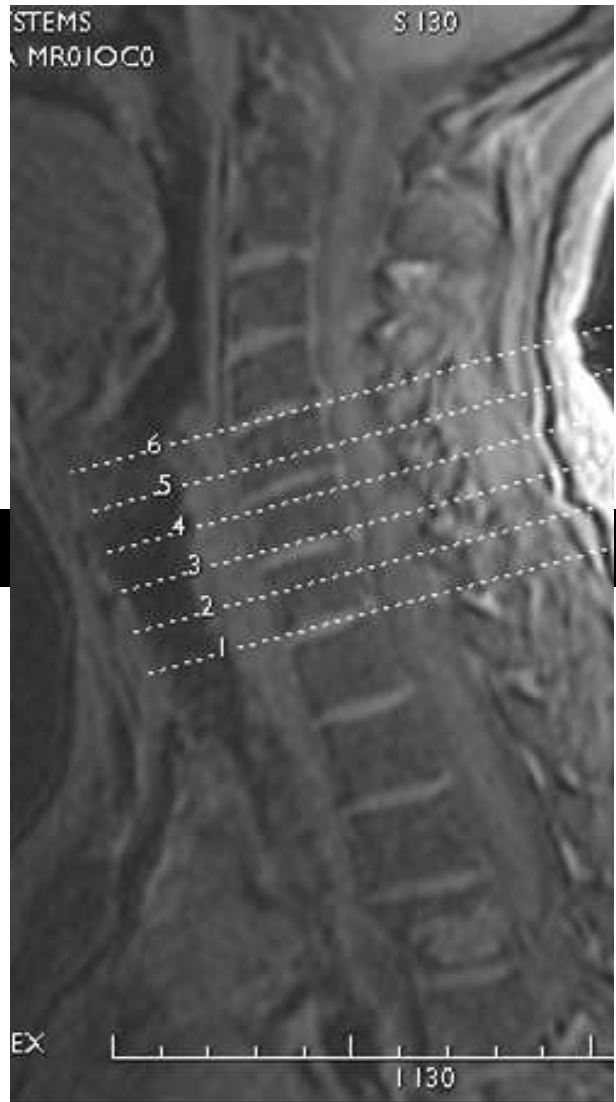
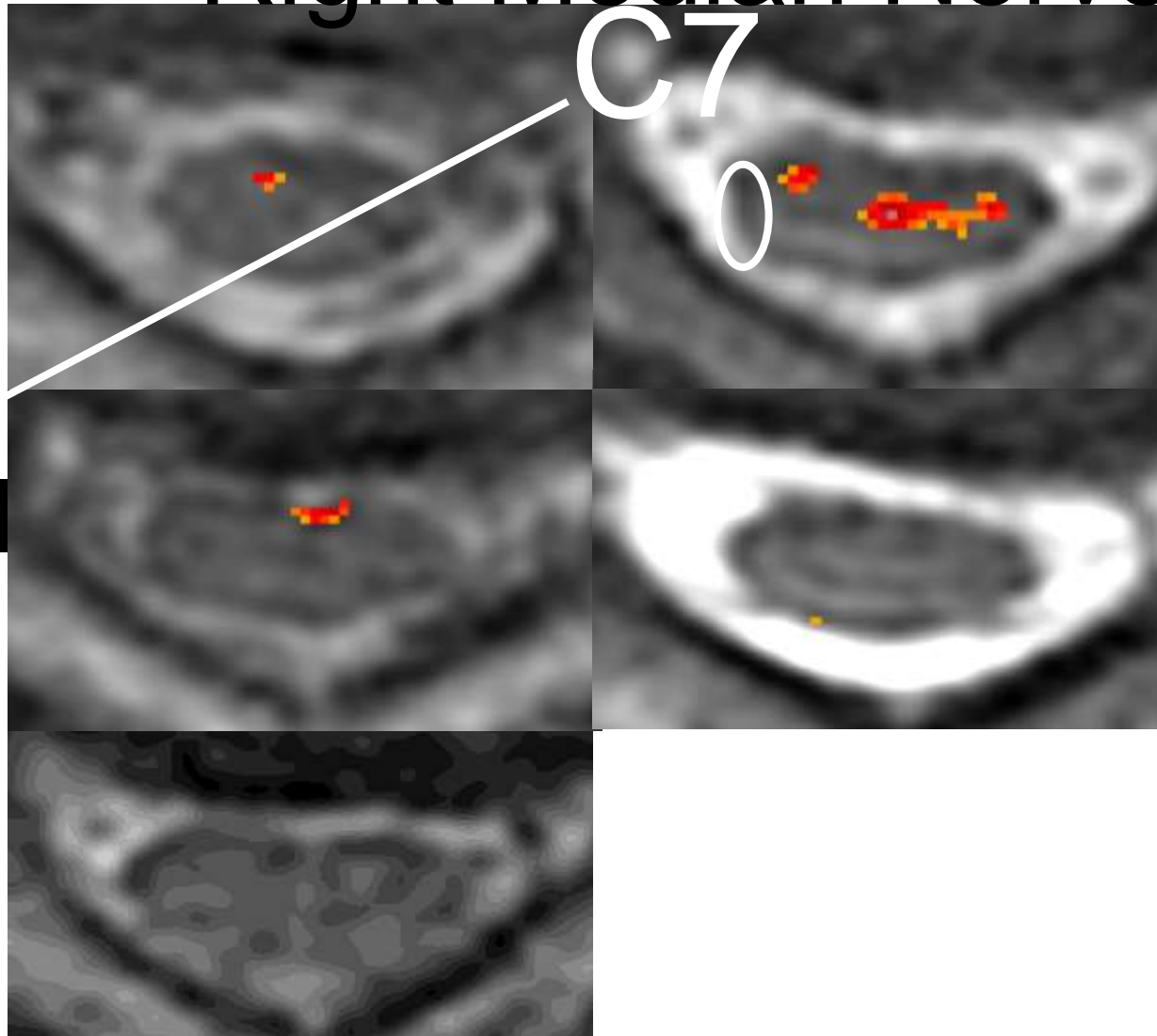
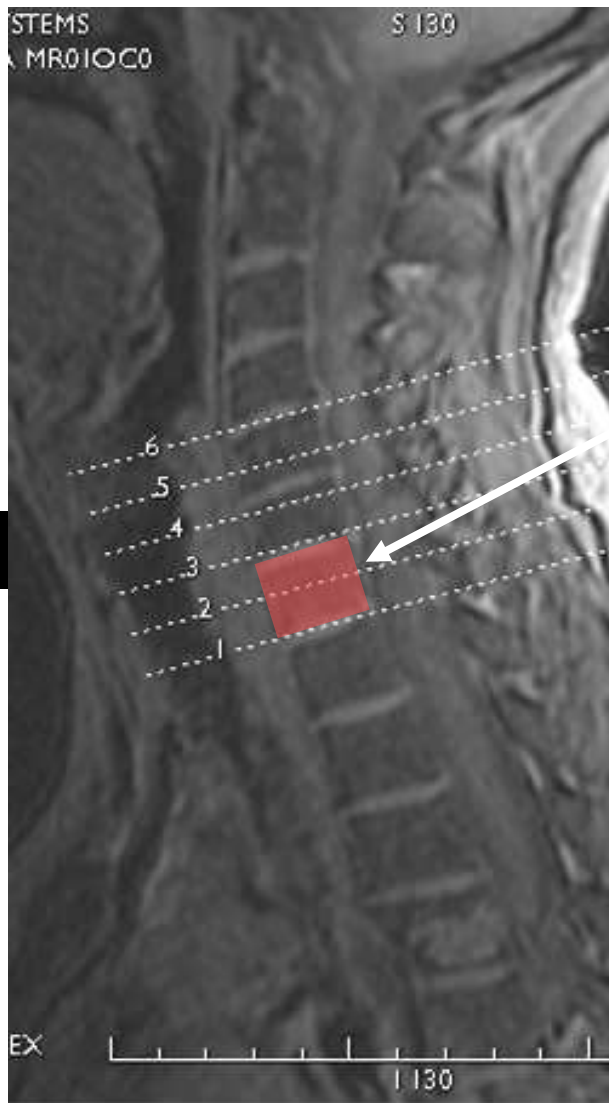
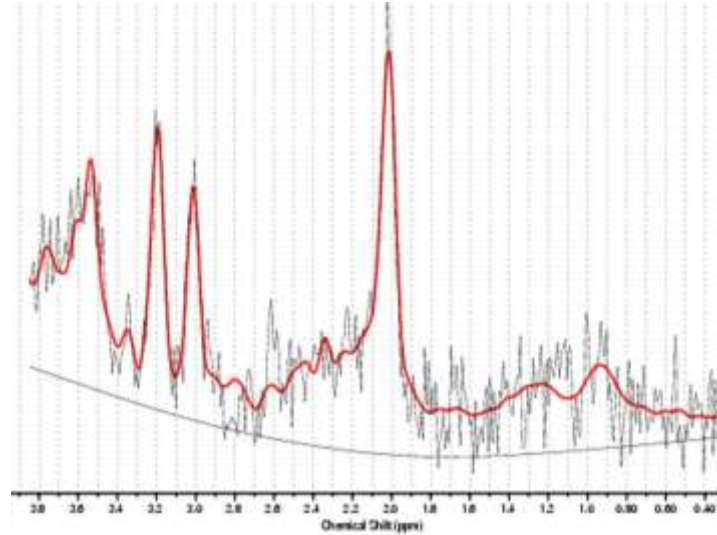
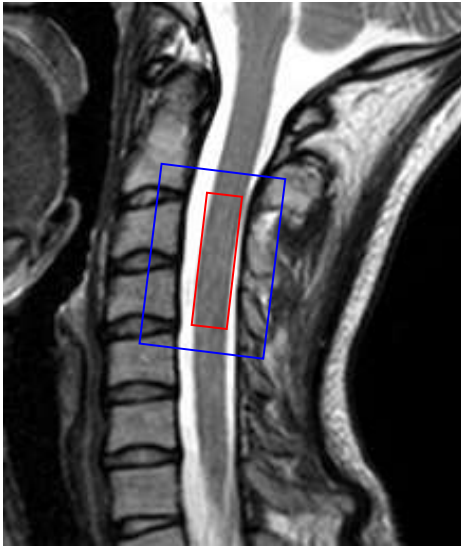
Gray Matter of the Spinal Cord

Rexed's laminae:

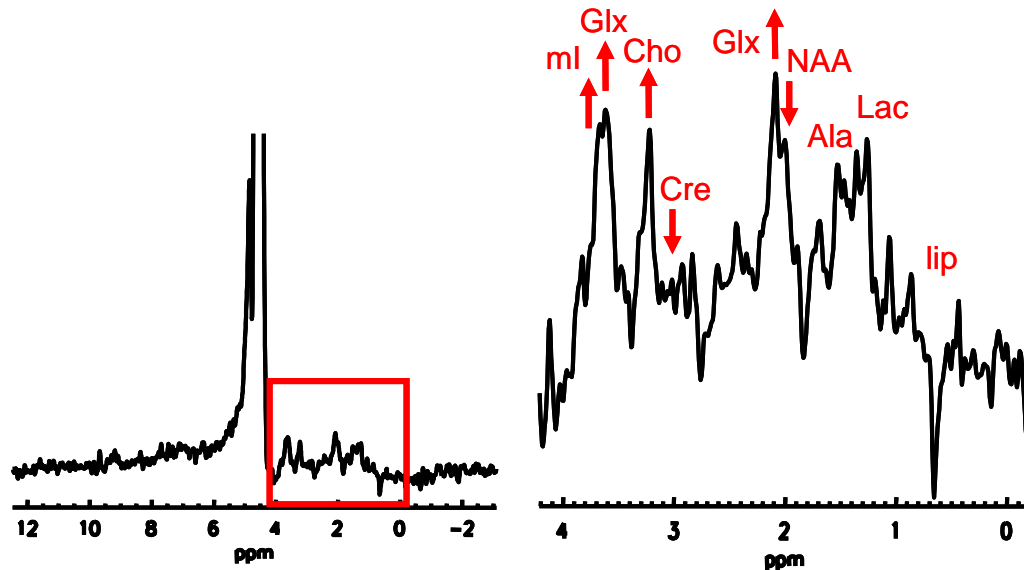
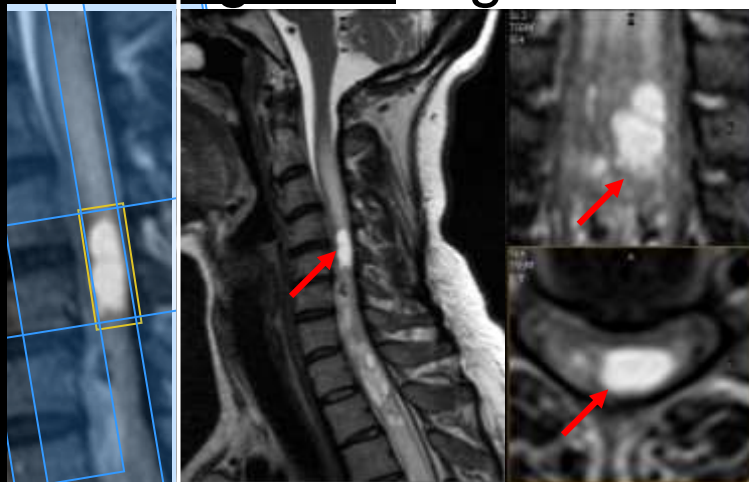
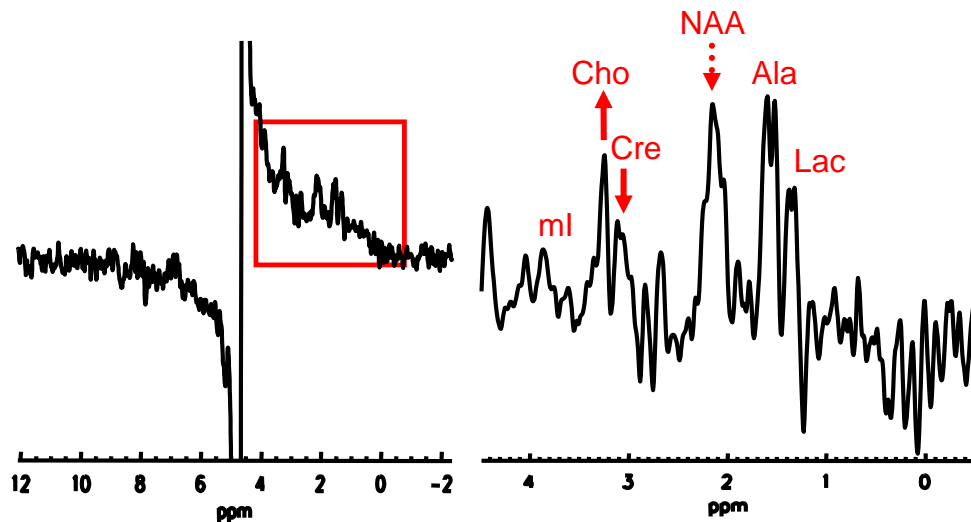
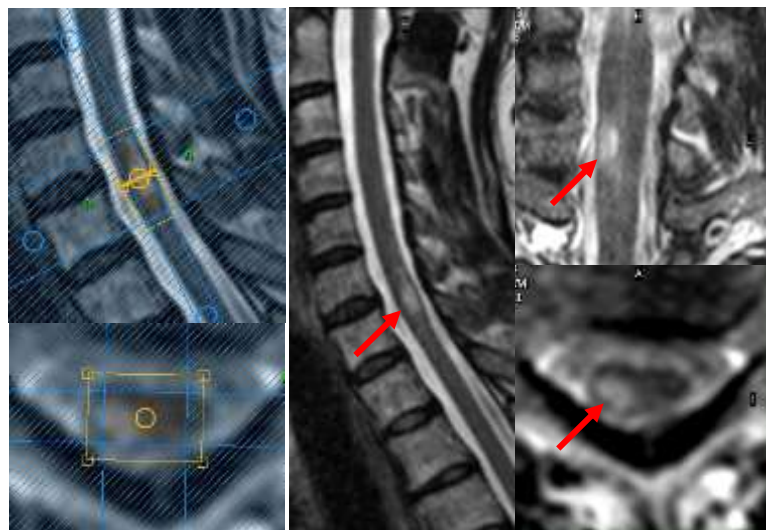
- Dorsal horn (I-VI)
  - projection neurons
- Ventral horn (VIII and IX)
  - Efferent neurons
- Intermediate zone (VII)
  - Interneurons (spinal reflexes)
- Region surrounding the central canal (lamina X)

Spinal cord fMRI: activation conditions

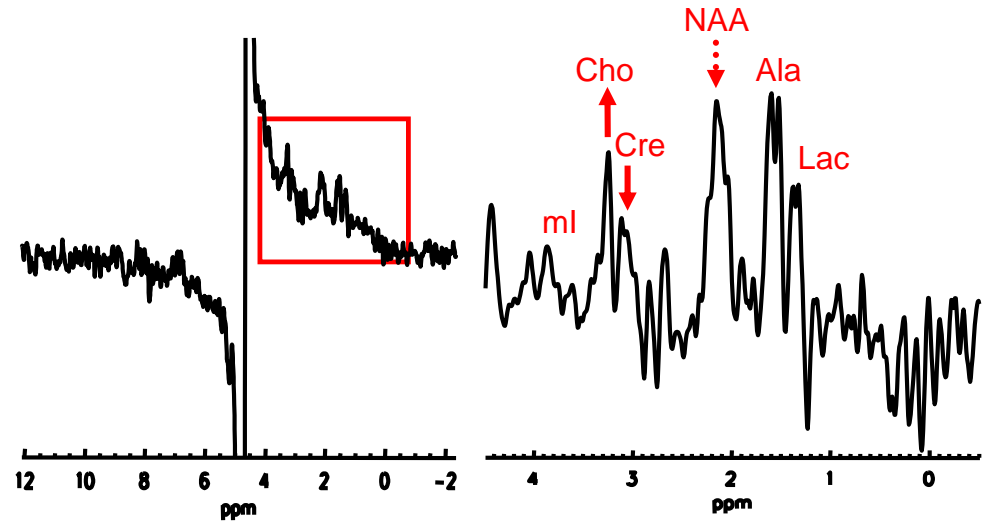
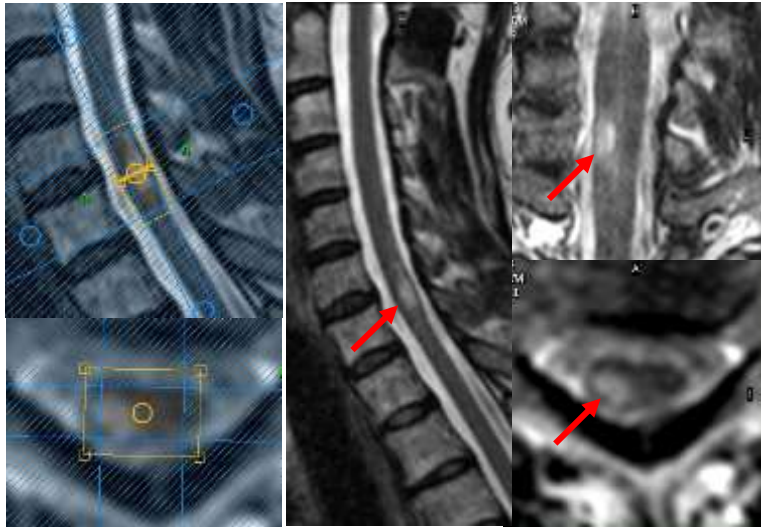
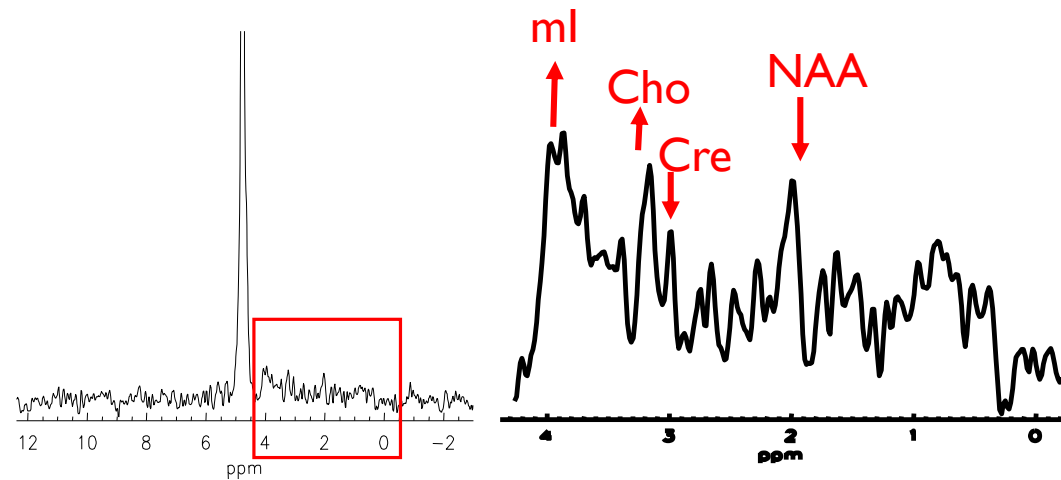
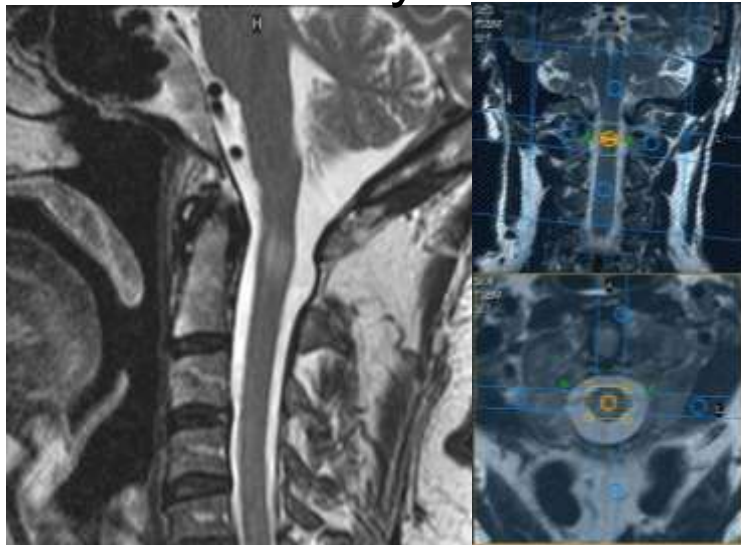
- Investigate whether the f-MRI signal can be spatially localized to particular anatomical locations specific to focal upper extremity motor tasks and sensory stimuli
  - motor tasks (R hand, R elbow)
  - sensory stimulation (thermal stimulus)
  - electrical stimulation

- motor activity was lateralized ipsilateral
- motor activity mainly in the intermediate and ventrolateral gray matter (Rexed's lamina VII & IX)
- comparison of hand and elbow shows the expected segmental organization of the SC

Right Median Nerve

S 130
MR01OC0
C7
6
5
4
3
2
1
EX
I 130

MR Spectroscopy of the Spinal Cord

Noninvasive information on the biochemistry of neuronal

Concentration changes of metabolites:

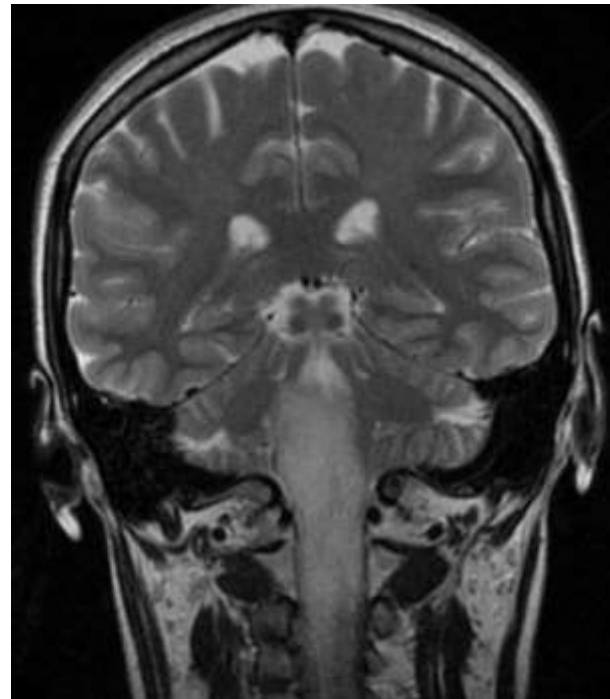
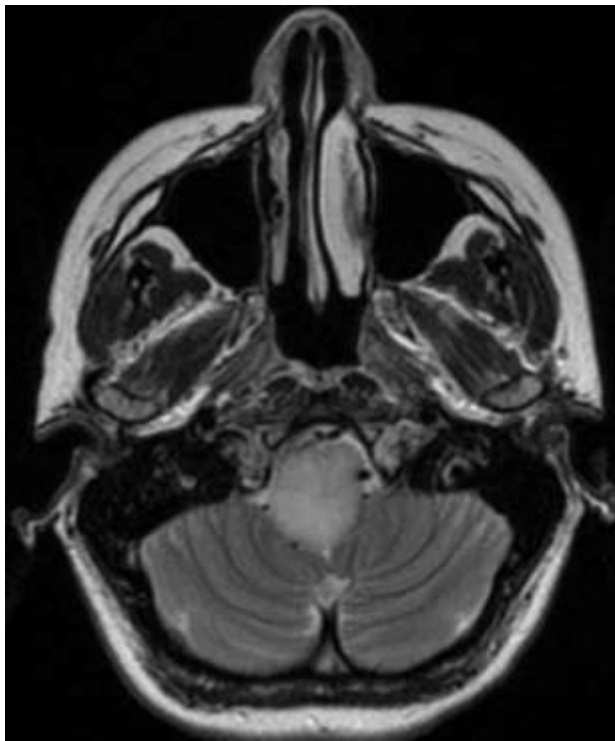
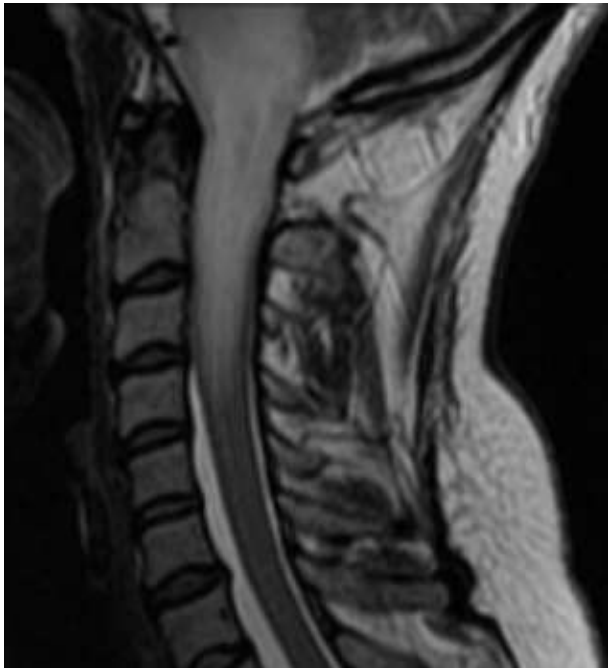
- N-acetyl-aspartate (NAA) → decreased neuronal densit
- Choline (Cho) → distorted membrane synthesis
- Creatine (Cre) → malfunction of the energy metabolism
- Myo-inositol (mI) → demyelination

"Quantitative magnetic resonance spectroscopy in the entire human spinal cord at 3T„ Magn Res Med (MRM) 59:1250–1258 (2008)

C 6/7 – low-grade glioma

C 1/2 – demyelination

"Quantitative magnetic resonance spectroscopy in the entire human spinal cord at 3T„ Magn Res Med (MRM) 59:1250–1258 (2008)

Tumori intramidollari

Terapia

- Priorità del trattamento è il controllo a lungo termine della lesione con preservazione delle funzioni neurologiche

- La chirurgia più efficace prevede la completa rimozione della lesione

Cavernomi spinali

- 1-3% di tutti i tumori intramidollari

- Jallo GI, Freed D, Zareck M, Epstein F, Kothbauer KF: Clinical presentation and optimal management for intramedullary cavernous malformations. Neurosurg Focus 2006; 21:E10.

Cavernomi spinali

- Analogamente ai cavernomi cerebrali sono lesione ben delimitate costituite da pacchetti di vasi simil capillari senza coinvolgimenti di tessuto nervoso

Cavernomi spinali

- Caratteritica deposizione di emosiderina ai bordi della lesione Ipointensa in T2

Cavernomi spinali

- Presentazione acuta
- Presentazione ricorrente
- Presentazione progressiva

Cavernomi spinali

- Microemorragie che portano a ialinizzazione e ispessimento, gliosi e alterazioni microcircolatorie. Talora parziale trombosi.

Cavernomi spinali storia naturale

Gross BA, Du R, Popp AJ, Day AL: Intramedullary spinal
cord cavernous malformations. Neurosurg Focus 2010 Sep; 29(3):E14.

- Overall annual hemorrage rate 2,5% (range 0-4,5%)

Cavernomi spinali
storia naturale

Sandalcioglu IE, Wiedemayer H, Gasser T, Asgari S, Engelhorn T, Stolke D: Intramedullary spinal cord cavernous malformations: clinical features and risk of hemorrhage. Neurosurg Rev. 2003 Oct;26(4):253-6.

- Prospective rehaemorrhage risk 66% per patient / year

chirurgia

- Indicata lesioni sintomatiche
- IOM

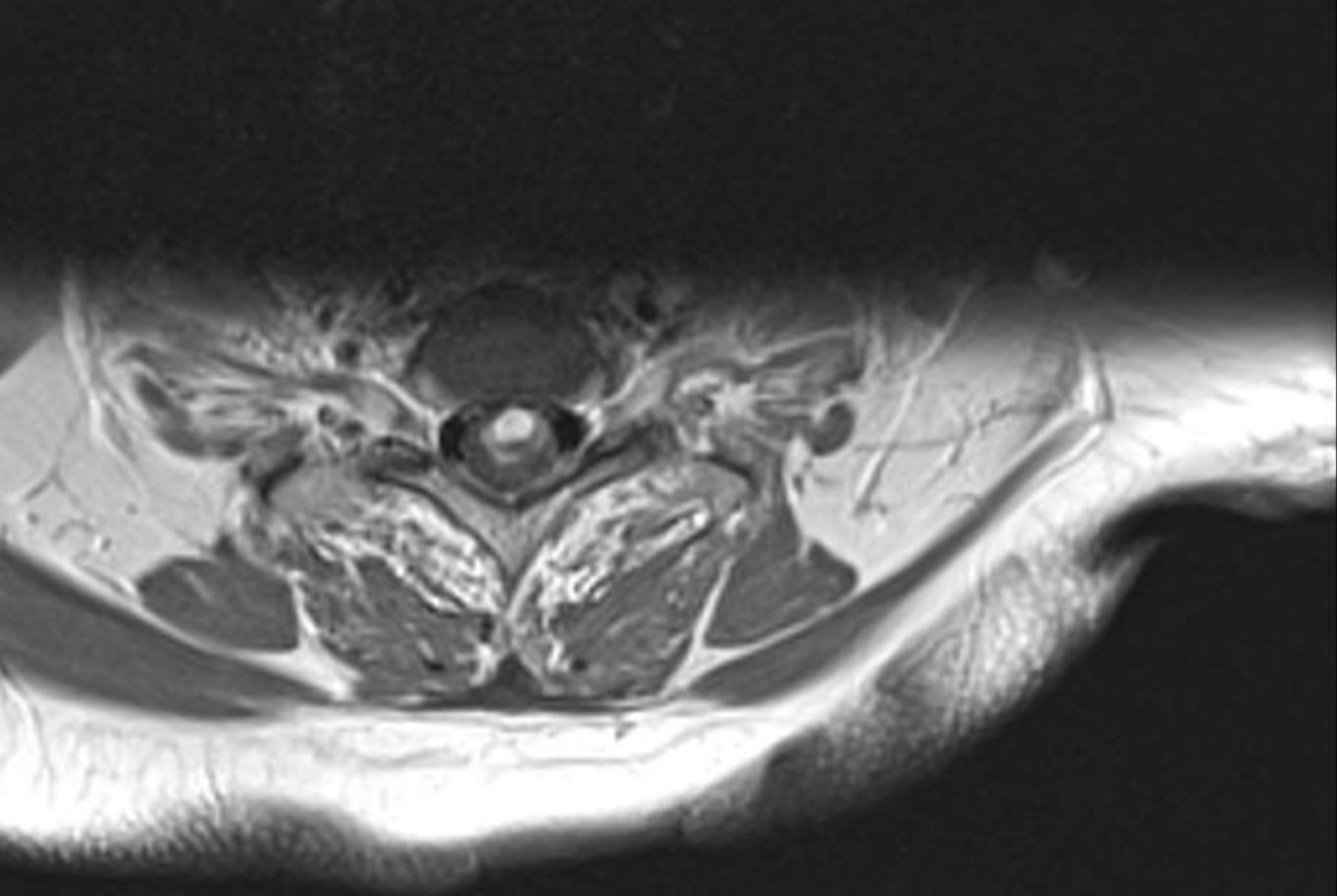
chirurgia

- Approccio posteriore nelle lesioni dorsali
- Approccio posterolaterle nelle lesioni ventrali(attne problema rotazione midollo)

Angevine PD, Kellner C, Haque M, McCormick PC: Surgical management of ventral intradural spinal lesions. J Neurosurg Spine. 2011 Jul;15(1):28-37.

- Donna 75 aa
- Esordio acuto di dolore urente nel braccio e regione scapolare sin. In due settimane progressiva perdita di destrezza nella mano e quindi evoluzione verso emiparesi e caduta della mano sin.

- Rm. A livello della giunzione cervicotoracica Lesione intramidollare ventrale disomogenea, ipointensa in T2

Diagnosi di cavernoma con esteso edema di tutto il midollo cervicale

- Review gruppo di studio francese

Labauge P. et al Surg Neurol 2008

Conclude: i cavernomi ventrali sono rari e con outcome peggiore

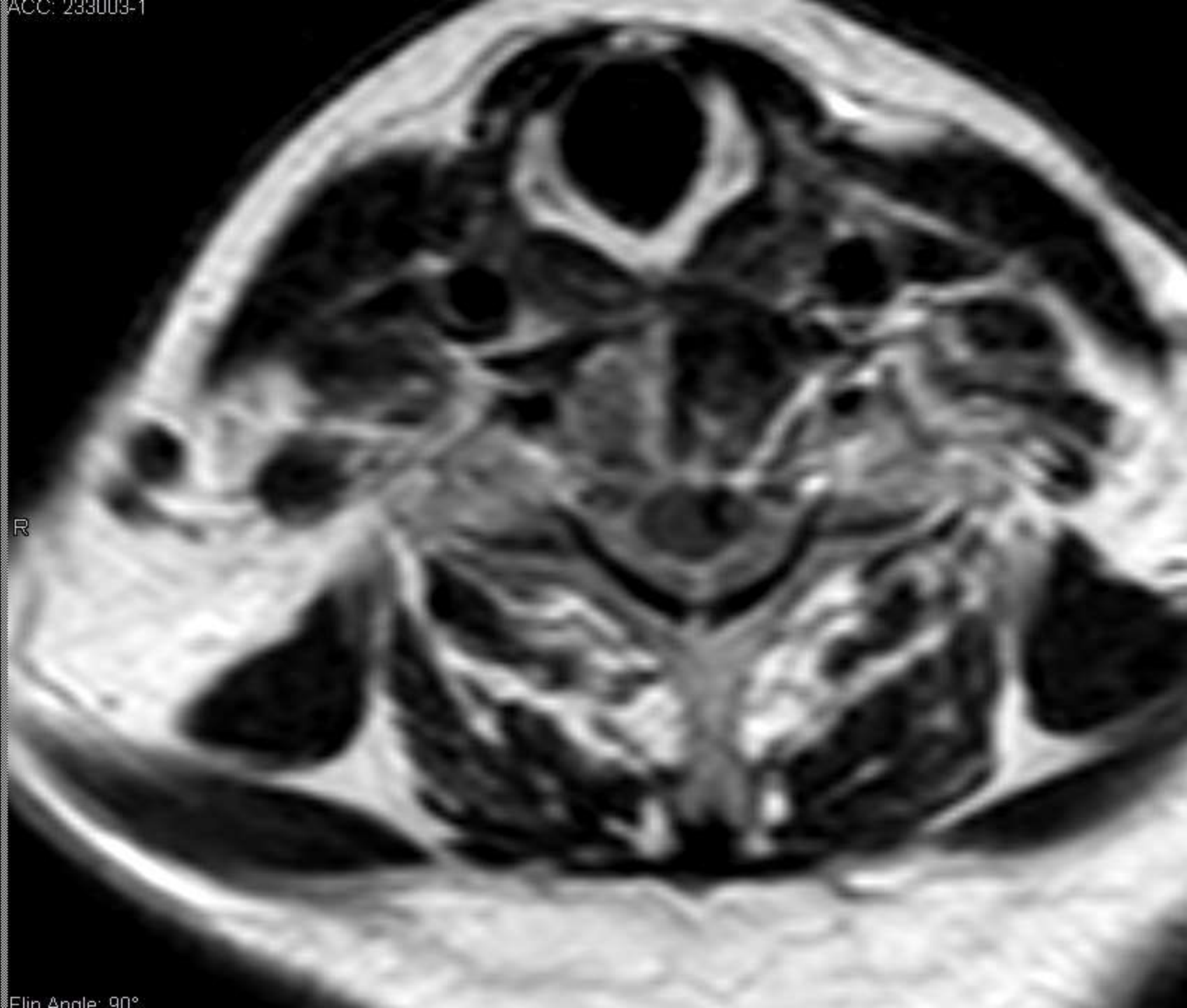
- Incisione laterocervicale lungo il bordo anteriore dello scm di sin e accesso al rachide per dissezione avascolare passando fra trachea ed esofago da un lato ed il fascio vascolonervoso. In visione microscopica discectomia C7-T1 e somatectomia C7-T1
- Apertura della dura sulla linea mediana, identificazione e rimozione del cavernoma con assistenza dl monitoraggio intraoperatorio

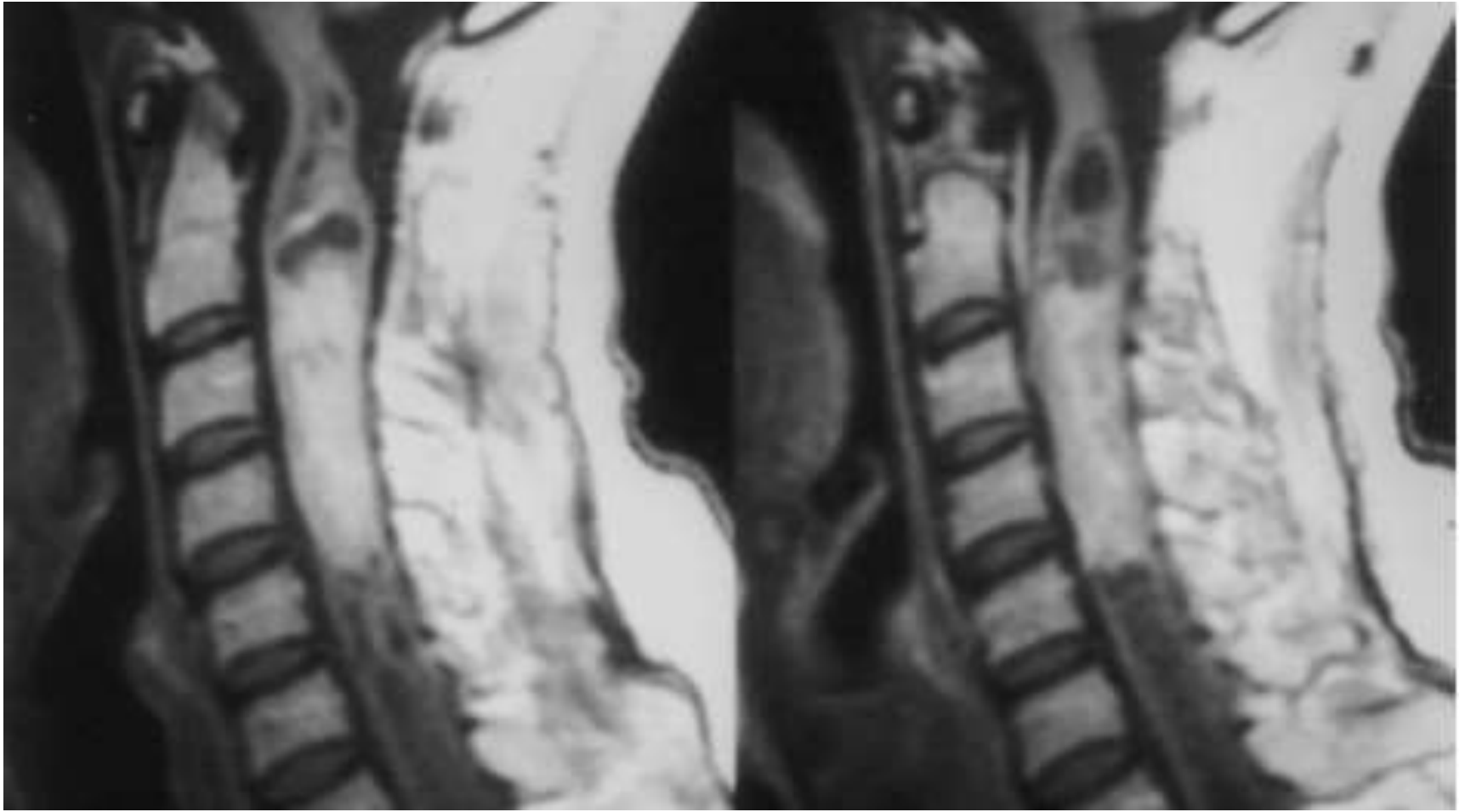
- Chiusura della dura con seta 4-0 e duralseal
- Mesh di titanio (pyramesh, medtronic riempita di un gel di osso sintetico) e placca di titanio (Zephir medtronic fissata con viti)
- Collare Phyladelphia 3 mesi

- Progressivo miglioramento dell’emiparesi enl follow up a 1-3-6 mesi e rm che dimostra la rimozione completa della lesione.

ACC: 233003-1
2.1
4.1
6.1
A
Flip Angle: 90°

2.1
4.1
6.1
A
Flip Angle: 90°

ACC: 233003-1
2.1
4.1
6.1
R

- In letteratura vi sono solo altri 7 casi analoghi

- George B, Zerah M, Lot G, Hurth M: Oblique transcorporeal approach to anteriorly located lesions in the cervical spinal canal. Acta Neurochir (Wien). 1993;121(3-4):187-90.

Conclusions

Aggressive surgery is the best choice most of the time.

-

IOM are mandatory for intramedullary tumors.

J Neurosurg Pediatrics 13:170–177

Quality of life after surgical treatment of primary intramedullary spinal cord tumors in children

Clinical article

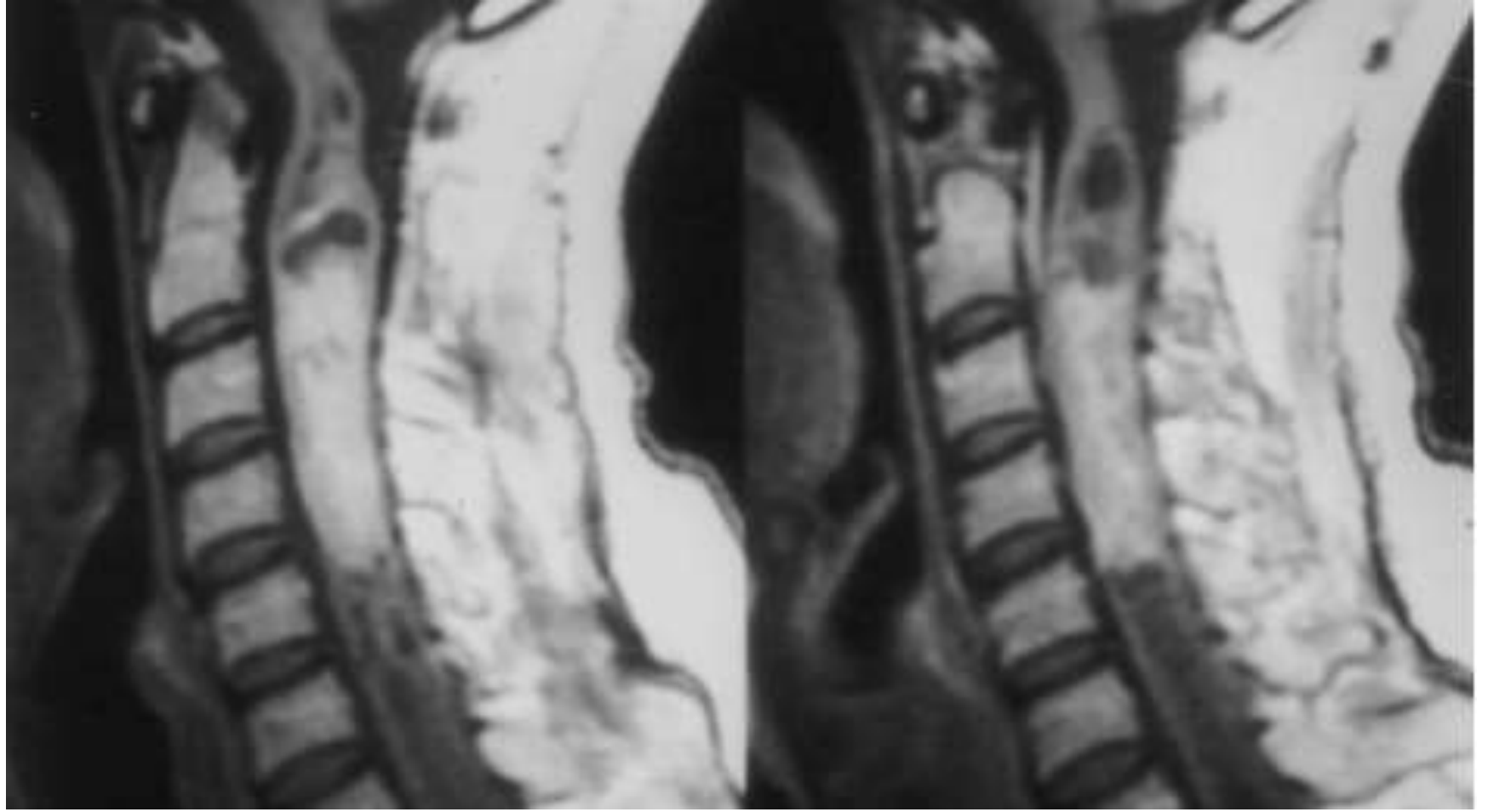
Christian Schneider, M.D.,[1] Eveline Teresa Hidalgo, M.D.,[1] Thomas Schmitt-Mechelke, M.D.,[2] and Karl F. Kothbauer, M.D.[1,3]

Divisions of [1]Neurosurgery and [2]Pediatric Neurology, Cantonal Hospital of Lucerne; and [3]Department of Neurosurgery, University of Basel, Switzerland

Subependymomas can rarely occur in the spinal cord, and account for about 2% of symptomatic spinal cord tumors. It most often occurs in the cervical spinal cord, followed by cervicothoracic junction, thoracic cord and conus medullaris. It often has an eccentric location in the spinal cord and lacks gadolinium enhancement on magnetic resonance imaging. We present a rare case of symptomatic subependymoma of the cervical spinal cord, which underwent successful gross total resection. Surgical pearls and nuances are discussed to help surgeons to avoid potential complications.

The video can be found here: **http://youtu.be/Rsm9KxZX7Yo.**

(http://thejns.org/doi/abs/10.3171/2014.V3.FOCUS14258)

Key Words • **cervical spine** • **intramedullary** • **spinal cord tumor** • **subependymoma** • **video**

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l tumor extends only from C2 to C5.
nd a large caudal polar cyst accoun
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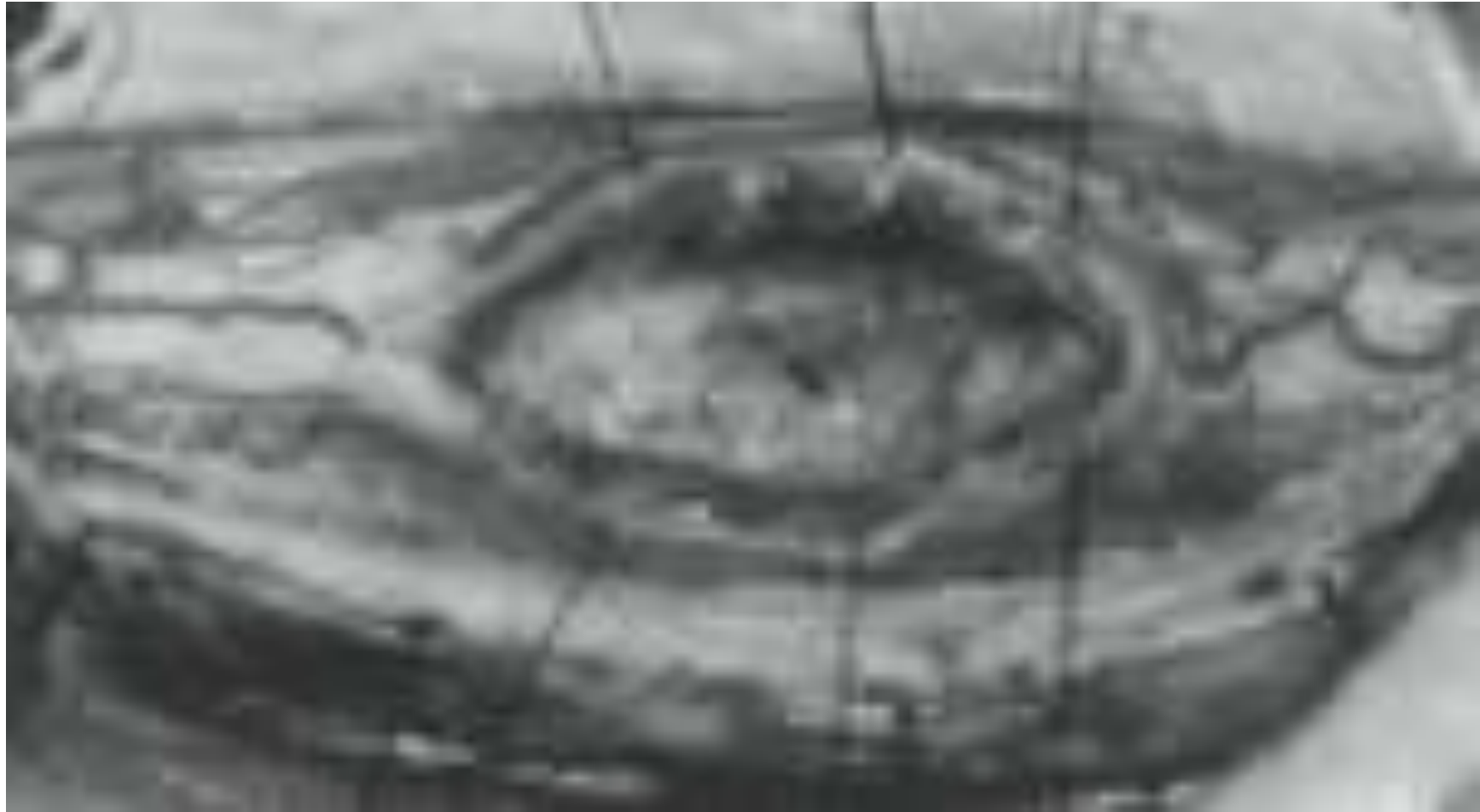
Fig. 1. Pre-operative (A) and post-operative (B) T2-sagittal spine MRI showing a patient (#17) with a loss of physiological cervical lordosis progressing to kyphosis after laminectomy.
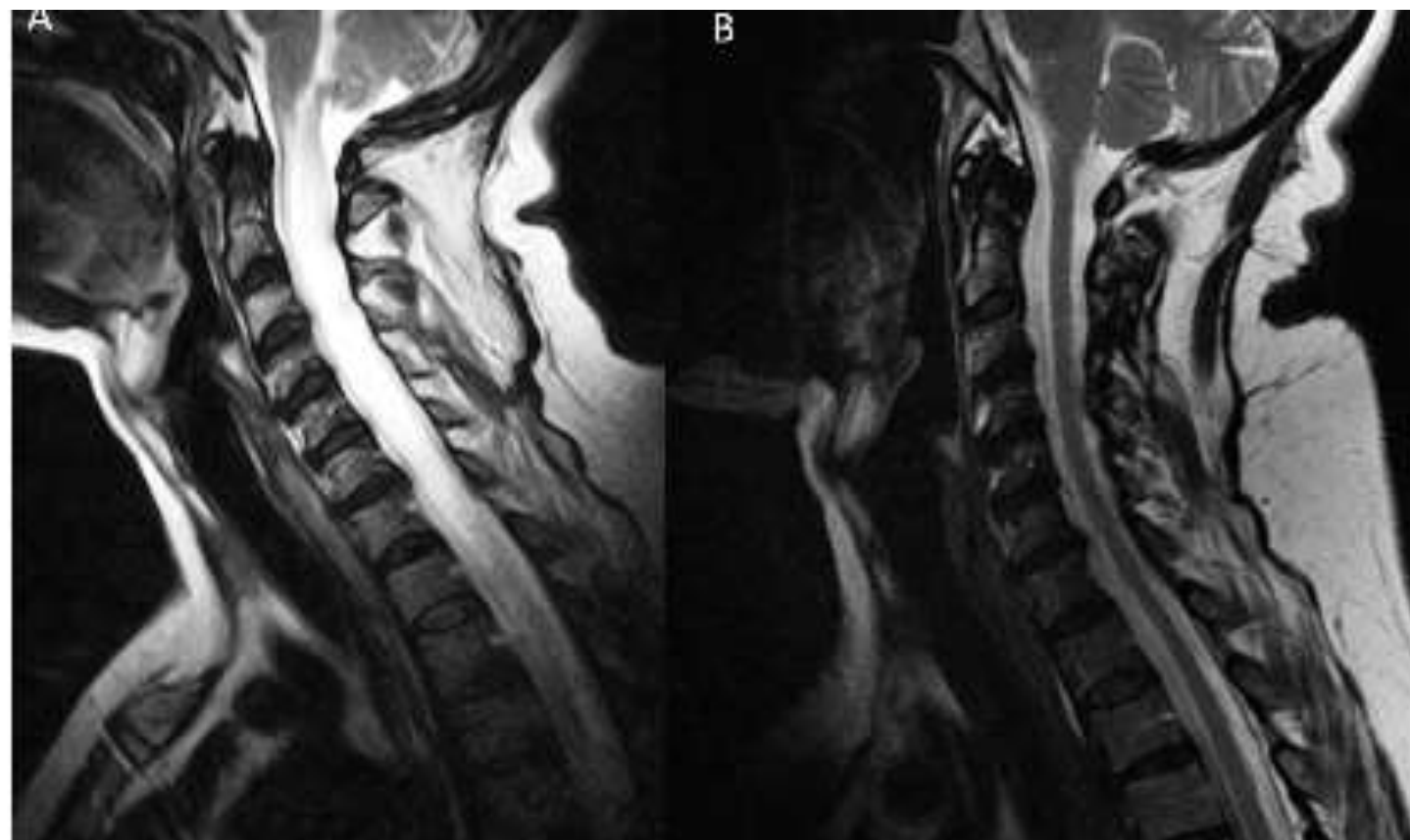
Fig. 2. Pre-operative (A) and post-operative (B) T2-sagittal spine MRI (#8) with no onset of deformity after cervical laminoplasty.

A
B